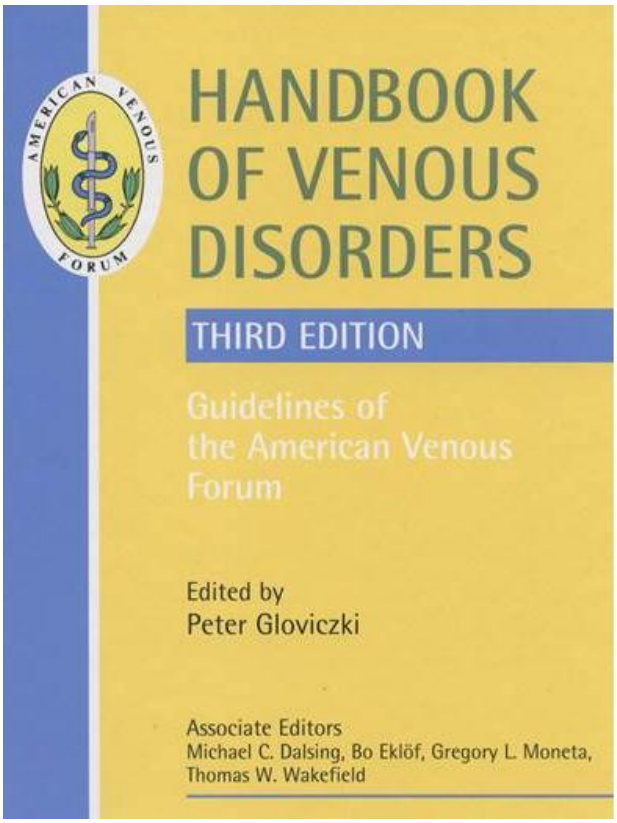
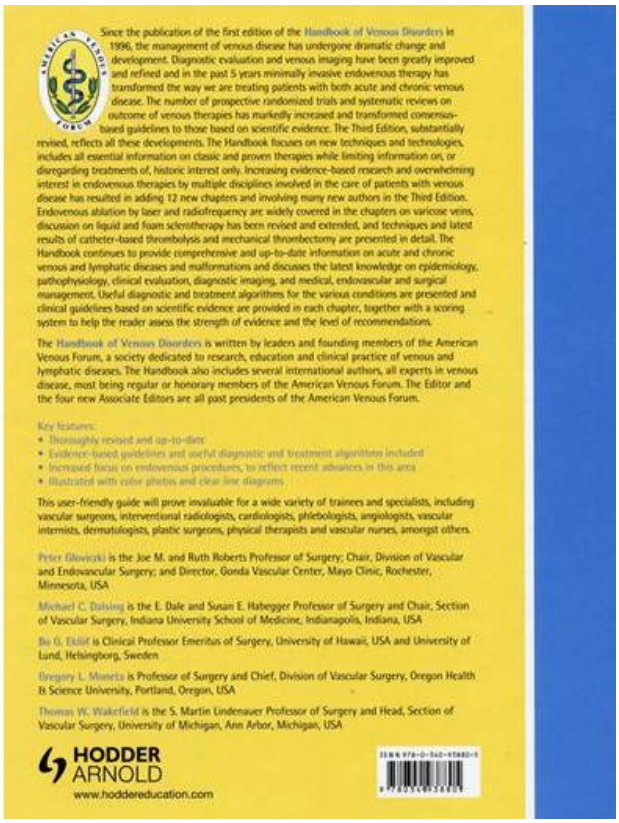
THE LAYMAN'S HANDBOOK OF VENOUS DISORDERS

Adapted from the Handbook of Venous Disorders; Guidelines of the American Venous Forum, Third Edition (Hodder Arnold, London 2009)





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An on-line book, **The Layman's Handbook of Venous Disorders**, designed to assist you with terms, treatment options, and answers to frequently asked questions.

Provided by the American Venous Forum: veinforum.org

The Editors of the Web-based publication: Dr. Michael C. Dalsing, MD and Gary W. Lemmon, MD (chapter adaptations by Kellie R. Brown, Teresa L. Carman, Raghu Motaganahalli, Gary W. Lemmon, Michael C. Dalsing)

The text of one of the most comprehensive textbooks on venous disorders has been adapted and rewritten in 20 chapters in layman's terms for easy understanding. The Editors of the Web-based publication, Dr. Michael C. Dalsing and Dr. Gary W. Lemmon, had two goals in mind: increase awareness of venous disease and educate the American public and our patients with acute and chronic venous disorders. The publication provides essential information on the cause, presentation and up-to-date management of venous diseases. Considerable new data are available on prevention and treatment of blood clots, leg swelling, varicose veins and venous ulcers.

The chapters, designed specifically for the web, are concise, easy to read, provide clear images, and each includes a section for frequently asked questions. Scan the Index or enter a key word, such as "varicose," in the search box above to instantly find answers to your questions.

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CHAPTER 1
NORMAL VENOUS CIRCULATION

Original author: Frank Padberg
Abstracted by Teresa L.Carman

Introduction

The circulatory system is responsible for circulating (moving) blood throughout the body. The heart and the blood vessels are the most important parts of the circulatory system. The heart is a central organ in the circulatory system. With each beat it forces blood into the blood vessels which transport or carry oxygen and nutrients to all of the tissues and organs (the arteries) of the body and then blood returns back to the heart through the veins.

There are three different types of blood vessels which play different roles within the circulatory system. The two main blood vessels are the **arteries** and the **veins**. The arteries carry the blood loaded with oxygen and nutrients away from the heart and the veins return the "used" blood, which has had the oxygen and nutrients removed, back to the heart. The **lymphatic vessels** are the third component. Briefly, they act as a "clean-up" system to pick up fluid, protein, and other debris left behind by the veins. They filter and clean the fluid before returning it to the heart.

Physiology/Hemodynamics

In normal circulation, the oxygenated blood leaves the left side of the heart through very large arteries. It flows through smaller and smaller **arteries** and even smaller **arterioles** and **capillaries** until it reaches the tissues and organs where the blood vessels are very small usually requiring a microscope to see them. This is called the "**capillary bed**". This is where the end of the arterial system connects to the beginning of the venous system. The blood vessels in the "**capillary bed**" are very tiny, thin walled vessels. This allows for easy release of oxygen and nutrients (sugars, fats, etc.) into the tissues **(Figure 1)**. Blood must then return through the **veins** to the right side of the heart where it can enter the lungs and pick up more oxygen. The venous system has **deep** and **superficial veins**. The **deep veins** are the major veins which return the blood. These lie within the muscles of the arms and legs. The **superficial veins** collect blood from the skin and are intended to take blood through the **communicating veins** back into the deep system. This is assisted by a series of one-way **valves**.

The regulation of **blood flow** through the blood vessels and to the tissues is under fairly complex control of the brain and nervous system as well as local chemicals which may be released by the tissues. Through out the course of the blood vessels there is a complex network of nerves that help regulate the flow through the arteries and veins. This allows tissues to have more blood flow when they are active or exercising and less flow when they are quiet or relaxed. For example, the blood vessels within the skin play a major role in maintaining body temperature. When it is cold they constrict or shut down and move blood away from the skin to the center of the body to preserve heat and when it is hot, they shunt more blood towards the skin to increase heat loss. Local injury or

trauma causes release of chemicals which may create either increased or decreased flow. Increased flow may be seen as localized swelling such as in the case of a burn or an ankle sprain.

The **veins** are fairly thin walled and are able to change their shape depending on the volume or amount of blood within the vein. The volume of blood is proportional or related to the pressure in the veins. When the amount of blood (or pressure) in the vein is low, the veins are flat like an empty balloon. As the volume (or pressure) increases, the vein expands, similar to an inflated balloon. If the pressures are elevated in the venous system and the veins are too full this may cause leaking into the tissues which is reflected as **swelling** or **edema**.

To normally circulate blood through the body there are 4 requirements **(Figure 2, a and b).**
(1) A pump – the **heart**
(2) A **pressure difference** or areas of high pressure and areas of low pressure
(3) A "**venous pump**" – the muscles
(4) A normal vein with intact **valves**

(1) The **heart** serves as the main pump in the circulatory system. Blood moves through the arterial system propelled by the force of the heart. Even at the level of the ankle the arterial blood pressure can be recorded with a blood pressure cuff. When the heart is not pumping well, as in heart failure, this will frequently cause **swelling**.

(2) A **pressure difference** exists between the legs and the right side of the heart where bloods returns to as it goes through the body. At the end of the capillary bed and moving into the venous system there is very little pressure in the veins and at the level of the heart there is even less venous pressure. This pressure difference helps move blood back to the heart. When you lay flat the blood can flow from the higher pressure in the legs to the lower pressure at the heart fairly easily. If there are higher pressures on the right side of the heart, from heart disease or lung disease, the normal pressure difference between the legs and the heart is changed and this may cause swelling.

(3) A **"venous pump**" - normal venous return requires a "pump", similar to the heart, to return blood from the legs to the heart. There are two muscular "pumps" in the legs. The main pump of the legs, responsible for generating most of the venous return (blood pushed back toward the heart), is the calf muscle. In the foot there is a network of veins which serve as a minor or secondary pump to "prime" the calf muscle pump of the legs. With each step the foot and calf muscles contract (squeeze) and forces blood up through the venous system against gravity on the way back to the right side of the heart. Loss of the normal **calf muscle pump** because of not walking normally (shuffling) or loss of normal ankle movement in a cast or splint or after a stroke will result in swelling of the leg because of a decrease in venous return. Some adults as they age their gait may change and they can develop a "shuffling" gait – this may cause swelling because of loss of the calf muscle pump.

(4) A **normal vein** with intact **valves** is required to control "one-way" flow in the veins. A normal vein with intact valves is also required for **normal venous return**. When the **calf muscle pump** contracts it lifts a column of blood up against gravity through the venous system. The venous valves act like steps on a ladder – supporting the column of blood until it reaches the right side of the heart and the pressure difference can pull the blood back to the heart **(Figure 2, a and b)**. A normal vein

is clean on the inside like a soda straw. If there has been **injury from a blood clot** and the inside of the vein is scarred or rough this will inhibit normal venous return. If the valves are not working properly, either because of prior injury or because they have worn out (are incompetent), they will not support the column of blood and cause more pressure in the veins this is called **reflux**. The weight of the blood pushing down in the veins causes increased pressure and can push fluid out of the veins into the tissues. This is one cause of **swelling** or **edema**, a main symptom of **abnormal venous function**.

If any of the above mechanisms of **venous return** are impaired for a short time – **swelling** or leaking of fluid into the tissues may be noted. This is commonly noted after a long flight or with short-term immobilization. When these mechanisms are chronically impaired and pressures are chronically or persistently elevated we can begin to see the effects reflected in the skin and tissues of the legs as long-standing swelling, **darkening** and **thickening of the skin**, and occasionally even **ulceration** or wound formation.

Conclusion

The **heart** pumps the blood to other parts of the body through tubes, **blood vessels**, called **arteries**. This arterial blood supplies blood rich in oxygen and food to very small blood vessel called **capillaries** that are in direct contact with your cells that need this nourishment. The used blood is taken back to the heart and lungs by **veins**. The veins must use the force of the heart, the force of the muscles in the leg and special one way **valves** to move the blood up the leg (especially when standing) and against gravity to make the return of blood to happen. The veins must be widely open to allow good movement of blood as well. When any part of this process fails; leg swelling, skin **damage** and **discoloration** and even skin breakdown (**ulcers**) can happen.

Commonly asked questions

Why do my legs swell?

Leg swelling may occur for many reasons. When it is related to **venous disease** it may be because of increased pressure in the **veins** from heart or lung disease or because the **veins** are not working properly because the **valves** are worn out or incompetent and there is **reflux** or backflow in the veins. Other **causes of swelling** may include liver disease, kidney disease, medications, problems with water balance, or mechanical loss of the “calf muscle pump”. Your doctor can do laboratory testing and vascular testing to determine what conditions you may have.

What if my swelling is only in one leg?

Swelling that affects only one leg may be more likely to be a problem with outflow or **obstruction of the vein**. You need to seek medical attention to make sure there is not an acute **deep vein thrombosis (DVT)** or other important cause.

What causes the veins to not work well?

In many cases, **veins** may wear out because of an inherited (related to the family) or genetic (your own gene make up) predisposition. If you have a family history of **varicose veins**, this may not be avoidable. **Trauma or injury to the vein** is another cause for the veins to not work well this is the case after a blood clot or **deep vein thrombosis**. **Increased pressure in the veins** from obesity may also contribute to the veins not working well.

What can I do about my swelling?
Once your doctors determine the cause, they can provide therapy to help with this problem. In many cases, **elevating the foot of the bed** to provide drainage of the leg at night and using **compression stockings** may be very helpful. Of course, if there is a problem with your heart or other organs your physician will need to address these concerns also.

Figure 1. Blood leaves the heart by large arteries (red) until it reaches the legs and arms and other organs by way of the peripheral arteries. The blood then goes through smaller and smaller blood vessels until it reaches the smallest arterioles. These are the smallest blood vessels used by the heart to pump oxygen and food to where it is needed. At the levels of the skin and other end organs, oxygen and food is given to the cells at the level of the capillaries (purple). After the oxygen and food is delivered blood returns to the heart through the veins (blue).

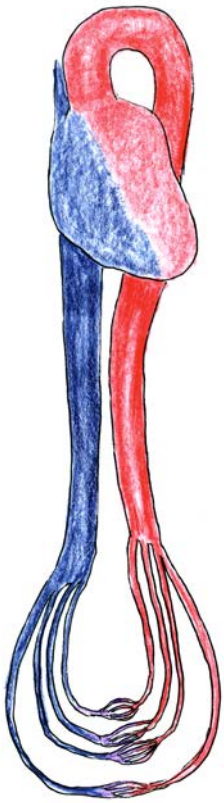
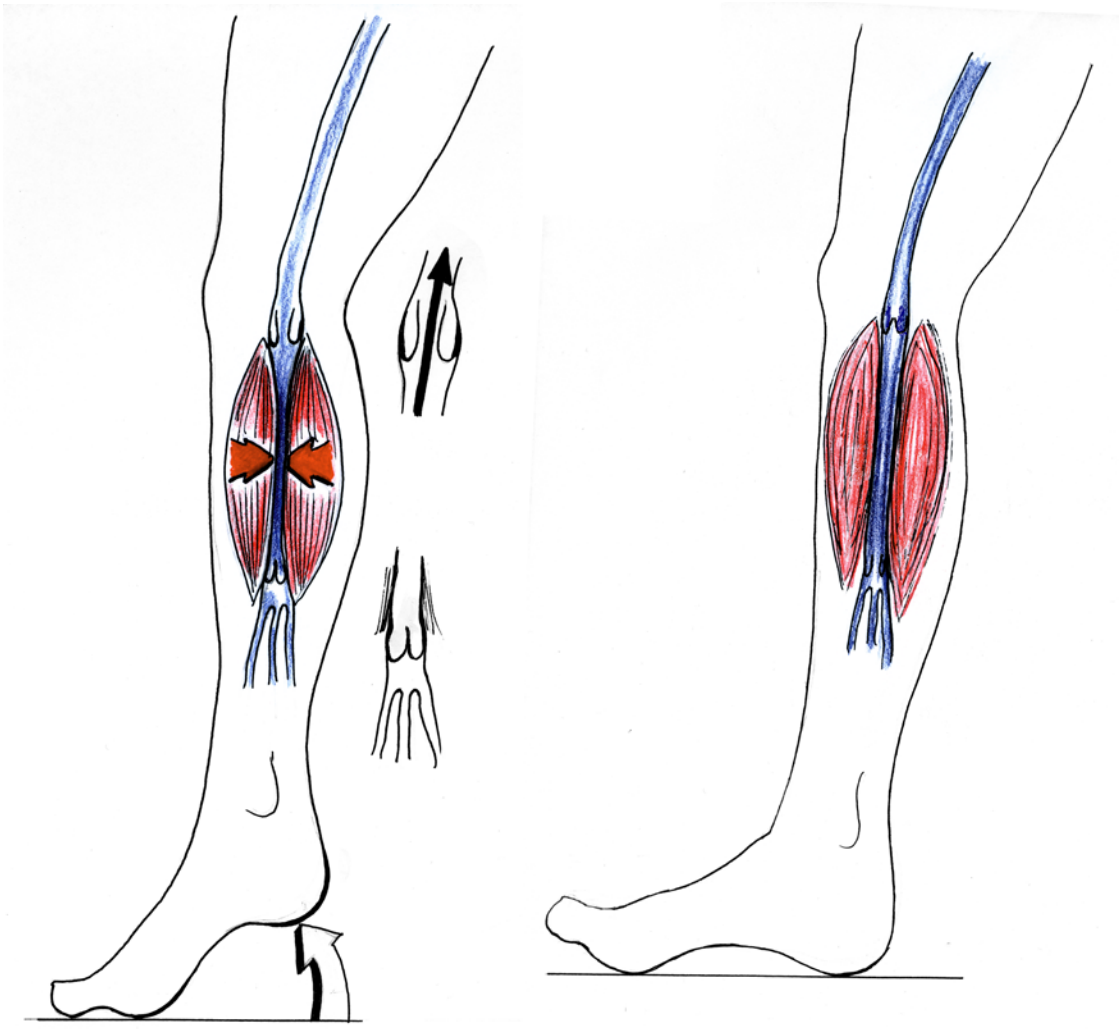


Figure 2. This artist's drawing shows how the normal calf muscle pump works to push the blood out of the leg and back to the heart and lungs. Venous flow is pulled from the superficial system (skin and fat under the skin) to the deep system (veins lying in the muscles) through a series of one-way valves. **(a)** When the calf muscle contracts this squeezing action forces blood forward against gravity and back towards the heart. **(b)** When the calf muscle relaxes the one-way valves closer to the heart close, preventing back-flow of venous blood.



(a) **(b)**

Provided by the American Venous Forum: veinforum.org

CHAPTER 2
RISK FACTORS FOR VENOUS THROMBOSIS

Original authors: Peter K. Henke, Mark H. Meissner and Thomas W.Wakefield
Abstracted by Kellie R. Brown

Introduction

Deep vein thrombosis (**DVT**, blood clots forming in the deep veins often of the leg, pelvic or abdomen but can also occur in the arm veins) is a serious healthcare problem in the US, with over 250,000 patients affected each year and at least 200,000 diagnosed with **pulmonary embolus** (**PE**). As our population ages, **DVT** is becoming more common. Among patients with **DVT**, one third of them are diagnosed due to a blood clot traveling in the blood vessels to the lung, causing shortness of breath and chest pain. This is called a **pulmonary embolus** (**PE**). The long-term effects of **DVT**, called **post-thrombotic syndrome** (**PTS**), affects about 500,000 patients with skin ulcers and millions more with discoloration and other skin changes in the legs. Because of the clot's ability to travel to the lungs, the effects of **post-thrombotic syndrome**, and the risk of recurrent **DVT**, it is important to prevent **DVT** from ever forming (prevention). In order to prevent **DVT** and **PE** some knowledge of who is at risk is needed. This chapter will discuss the common risk factors (those things that increase the bloods tendency to clot) for **DVT**, the changes that occur in a vein after a clot has formed, and what happens to the clot over time.

How common are DVT and PE?

Because many **DVT**s don't cause symptoms, knowing the true incidence is difficult. However, most sources believe that first time, symptomatic **DVT** or **PE** (those patient's who have symptoms from the clotting) in the USA happens in between 71 and 117 cases per 100,000 people. There are approximately 275,000 new **VTE** cases per year in the US, a figure that has not changed in the past 10 years. **VTE** stands for venous thromboembolism, which includes both **DVT** and **PE**.

What causes DVT and PE (VTE)?

There are many causes of **VTE**, including genetic risk factors and environmental risk factors. Some risk factors are permanent, and some, like major surgery, are temporary. Most risk factors have to do with a change in the blood's clotting ability, either due to different factors in the blood, or a change in a patient's activity level. Well established risk factors for **DVT** are discussed below. The development of **DVT** or **PE** most often occurs when a person has more than one risk factor.

What are the risk factors for VTE?

- **Age, Gender and Race**

Age, gender, and race may potentially influence the risk of **VTE**. Among these, age has most consistently been associated with increased **VTE** risk. As **age** increases, so does the risk. This increase in risk appears to be related to several age related factors, including decreased mobility, an increased number of other major risk factors such as cancer, age related changes in the blood's tendency to clot, and changes in the veins themselves.

Gender differences are less clear, with the existing studies differing on whether gender influences risk or not. The lack of consistent data makes it unlikely that there are major gender differences in the incidence of **DVT**. However, the risk in women is higher during childbearing years, whereas between the ages of 45 and 60 the risk may be higher in men.

Race has a variable affect on incidence of thromboembolism. There is evidence to suggest that the occurrence of post-operative **DVT** is less in Asian, Arab, Hispanic, and African populations than among those of European descent. This difference is probably due to genetic factors associated with **VTE**.

- **Surgery**

The increased risk of **DVT** or **PE** associated with surgery is due to decreased ability to move after surgery, and changes in the levels of certain clotting factors in the blood that occur after major surgery. The degree of risk also varies with the age of the patient, the length of the operation, the type of operation, and the presence of other **DVT** risk factors. Without treatment, **DVT** can occur in about 25% of patients undergoing general surgery, and can be up to 50% in patients undergoing hip or knee surgery. This risk can be greatly decreased with the appropriate use of sequential compression devices on the legs and/or doses of blood thinners before and after surgery.

- **Trauma**

A person who has had a significant trauma (in a car crash, for example) is at great risk for **DVT** and **PE**. These patients are generally unable to move for long periods of time due to their injuries. Often they are severely ill and are unable to have blood thinners due to their injuries.

- **Medical Illness**

About 60% of **VTE** events are related to confinement to a hospital or nursing home. Medical conditions that carry a high risk for **VTE** in hospitalized patients include congestive heart failure, major respiratory illnesses, cancer, sepsis, inflammatory bowel disease, and acute neurologic disease (such as stroke).

Malignancy (cancer) carries a significant risk for **VTE**. Up to 30% of patients with cancer can develop a **DVT**. While any cancer can increase the risk of **DVT**, leukemias and lymphomas have a particular risk, as do lung and gastrointestinal cancers. The risk

of **DVT** is highest early after the diagnosis of cancer, and cancer can sometimes be diagnosed in patients who present with a **DVT** for initially unknown reasons.

- **Immobilization or Travel**

Bed rest or other immobilization is also associated with an increased risk of **VTE**. This has to do with blood "pooling" in the **veins** for long periods of time, a condition called "stasis". Stasis of blood in the veins is the reason that people on long air flights or long drives are thought to have increased risk of **VTE**. This risk can mostly be erased by occasionally moving about during the travel and using knee-high elastic compression stockings. Older age, obesity, previous history of **VTE**, use of oral contraceptives and underlying blood clotting disorder increase the risk of travel-related clots.

- **Primary Blood Clotting Disorders**

There are several known genetic conditions that alter the presence or amount of **clotting factors** in the blood. These conditions often are associated with an increased risk of **DVT** or **PE**, but usually cause a problem only when another risk factor is also present. These existing genetic conditions include **antithrombin III (AT III) deficiency**, **protein C deficiency**, **protein S deficiency**, **Factor V Leiden mutation**, **anti-phospholipid antibody syndrome**, **prothrombin 20210A gene mutation**, **hyperhomocysteinemia**, **alterations in plasmin generation** and **increase in factors II, VIII, IX or XI**. The amount that each of these conditions affects **DVT** risk varies with the condition and with whether the patient has either one or two genes coding for the defect.

- **Oral Contraceptives and Hormonal Therapy**

Estrogen (female hormone) replacement, either in oral contraceptives (birth control) or in hormone replacement therapy, has been associated with an increased risk of **VTE**. The higher the estrogen amount, the higher the risk. Although it is less clear, progestin components in some contraceptives may also be related to an increased risk of **VTE**. The risk for **VTE** in women who take oral contraceptives is greater in women over 40, and in smokers. In general, oral contraceptives are discouraged in smoking women over 40, or in women with one of the genetic clotting disorders.

Although, the doses of estrogen used for post-menopausal replacement are about 1/6 of those used for contraception, there is a 2-4 fold increase in the risk of **VTE** in women who use them. However, the risk of replacement therapy must be kept in perspective, as it causes only about 2 new cases of **VTE** per 10,000 women per year.

- **Pregnancy**

There is an increased risk of **DVT** and **PE** in pregnancy, primarily due to a combination of changes in the blood clotting factors and compression of the pelvic veins by the fetus. There is also a risk of **VTE** after delivery. Once a mother has had a **DVT** during pregnancy, she is at increased risk for a recurrent **DVT** during subsequent pregnancies.

How does clot affect the vein?

After clot forms in a **vein**, **inflammation** occurs in the vein wall and in the clot itself. This **inflammation** leads to extension of the initial clot and clot organization. Over several weeks to months the clot will usually recanalize. **Recanalization** is a re-opening of the vein that was previously blocked by clot. This organization and re-opening of the **vein** causes damage to the **vein wall** and the **vein valves**. **Recanalization** does not always happen completely. The body has very complex pro-inflammatory and anti-inflammatory response systems. The extent to which these systems are activated and how they interact may have a role in how soon or how well the clot is organized and recanalized.

Does the vein return to normal after the clot resolves?

The answer to this question is generally no. The normal **vein wall** is very elastic and thin. It can change size to a great degree. After a **DVT**, the vein wall becomes stiff. This is due to breakdown of the **elastin** and **collagen fibers** in the vein wall. In addition to these changes, the vein wall also becomes fibrotic over time after a **DVT**. **Fibrosis** is a process of "toughening" of tissue, similar to what is seen in scar formation. This **fibrosis** can also affect the **vein valves** causing them to stop functioning properly. This can all lead to **veins** that are stiff, tough and scarred, with **valves** that don't function properly, and to increased pooling of blood in the **vein**. These changes in the **vein wall** and the **vein valves** contribute to the **symptoms** experienced in patients long-term after **DVT**, which include **leg pain, aching, heaviness, fatigue, and swelling**. This collection of **symptoms** is referred to as **post-thrombotic syndrome (PTS)**. While not everyone gets **PTS**, wearing **compression stockings** for at least 2 years after getting a **DVT** is thought to prevent or decrease the severity of **PTS**.

Conclusion

DVT and **PE** are common conditions. The **risk factors** include both genetic and environmental factors. **DVT** and **PE** generally occur when 2 or more of these risk factors occur at the same time. Decreasing the risk of **DVT** and its consequences involves decreasing immobility, using appropriate **anticoagulation**, and wearing **compression stockings** when appropriate.

CHAPTER 3
CLOTTING DISORDERS

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Abstracted by Kellie R. Brown

Introduction

The normal balance between **clot formation** and breakdown can be changed by the presence of certain genetic or acquired defects leading to abnormal clot formation. Reasons for the **clot formation** and breakdown processes to be unbalanced toward abnormal clot formation include **blood vessel injury**, **venous stasis** (lack of movement of the blood in the veins), and **clotting disorders**. These three factors make up **Virchow's triad**. An alteration in any one of these three factors can lead to abnormal clotting. All **risk factors** for **DVT** or **PE** fall into one of these three categories. A **venous thromboembolic** event (**VTE**) is either a **DVT** or **PE** or both in the same patient.

Clotting disorders are present in the majority of patients who have a **DVT**. The typical story for a person with an inherited **clotting disorder** is a spontaneous **DVT** at an early age. Doctors now have a variety of tests that can be done to test for an inherited **clotting disorder**. There is controversy over which patients should get which tests and what positive results mean. This chapter reviews the most common **clotting disorders**.

What is a D-Dimer Level?

D-dimer is a by-product of clot breakdown. The **D-dimer** level is not really a risk factor for getting a **DVT** or **PE**, but it may be elevated when a **DVT** or **PE** is present. The **D-dimer** level is used to "rule out" (show it is not present) **DVT** when the suspicion for **DVT** is low. This is because if the level is normal, it is unlikely that a **DVT** exists. However, if the level is high, **DVT** may or may not be present. Elevated **D-dimer** levels can indicate the presence of abnormal clot, but levels can also be elevated from other causes such as recent surgery, bleeding, trauma, pregnancy, cancer or abnormal blood clot in an artery.

Clotting Disorders

There are 2 types of **clotting disorders**. The first is a **hereditary disorder** that is inherited from one or both parents. The second is an **acquired disorder**, which a person is not born with, but that develops later in life.

Hereditary Clotting Disorders

The hereditary clotting disorders come in 2 groups:
Group 1: A lack of anti-clotting factors in the blood
Group 2: An increased amount of pro-clotting factors in the blood

Group 1 disorders include **anti-thrombin deficiency**, **protein C deficiency**, and **protein S deficiency**. **Group 2 disorders** include **activated protein C resistance** (**Factor V Leiden mutation**), **prothrombin G20210A mutation**, and elevated levels of **Factors VIII, IX, and XI**. In general, the Group 1 disorders are less common but more likely to cause abnormal clotting than Group 2 disorders. Patients with Group 1 disorders usually have their first **DVT** at a young age, have a higher likelihood of recurrence, and are more likely to have a family history of **DVT** or **PE** than patients with a Group 2 disorder.

GROUP 1 DISORDERS:

A LACK OF ANTI-CLOTTING FACTORS IN THE BLOOD

Antithrombin Deficiency

Antithrombin is a natural blood thinner found in the body. It works to reduce clot formation. Over 100 gene mutations have been found that can lead to antithrombin deficiency. This disorder is inherited as an **autosomal dominant trait**, which means that if a person gets an abnormal gene from one parent and a normal gene from the other parent, they will have the disease.

How common is it?
Antithrombin deficiency is present in 0.07-0.2% of the general population and 0.5-8% of those with **DVT**.

How and when do you test for it?
The amount of antithrombin in the blood can be tested with a blood test.

Tests should be drawn 3 months after the **VTE** happens, and at least 5 days after any **blood thinner** has been stopped. This is because **blood thinners** affect the levels of **antithrombin** in the blood. Many other conditions can also affect the **antithrombin** levels, so test results should be interpreted with the patient's entire medical history taken into account.

What is the risk of VTE in a person with antithrombin deficiency?
Antithrombin deficiency is a strong risk factor for **DVT**. The risk in most people with **antithrombin deficiency** is increased by 5 to 50 times. Most patients with this disorder will have had a **DVT** by age 30. Abnormal clotting in the arteries has been reported in people with this deficiency but it is uncommon and its association with the deficiency is not clear.

How do you treat antithrombin deficiency?
Patients with **antithrombin deficiency** are resistant to **heparin** therapy because **heparin** requires the presence of **antithrombin** to work. **Heparin** is a commonly used

intravenous **blood thinner**. Patients with **antithrombin deficiency** who need **blood thinners** should get another type of **blood thinner** that does not need **antithrombin** to work. **Antithrombin** itself can be given if necessary. After a **VTE** has occurred, lifelong oral **anticoagulation** is generally recommended.

Protein C Deficiency

Protein C is a natural **anticoagulant** that is made primarily in the liver. During the clotting process, **protein C** is activated, and along with **protein S** acts as a **blood thinner** to keep the clotting process in check. **Deficiency in protein C** results in decreased ability to keep the clotting process in check, leading to abnormal clot formation.

How common is it?

Protein C deficiency is present in up to 0.4% of the population and may be present in about 4% of patients with **DVT** or **PE**.

How and when do you test for it?

The amount of **protein C** in the blood can be tested for with a blood test. The test should be done 2-4 weeks after any **warfarin** therapy is stopped. Many things can cause **protein C** to be low, such as new clot formation, low **Vitamin K**, liver disease, severe infections (sepsis), kidney failure, post-operative state, breast cancer patients after certain chemotherapies, and massive bleeding.

A normal **protein C** level after a new clot has occurred rules out the disease, but a low level in this situation would need to be re-checked after therapy for the new clot is completed before the diagnosis could be made.

What is the risk of VTE in a person with protein C deficiency?

People with **protein C deficiency** are about 3 times more likely to experience a **DVT** or **PE** than the general population. By age 40, about 50% of those with **protein C deficiency** will have had a **DVT** or **PE**. There is not a significantly increased risk of artery clot in these patients.

How do you treat protein C deficiency?

If a person has **protein C deficiency**, and they have never had a **VTE**, then no medication is required. They need to have **blood thinners** given to prevent **VTE** prior to surgery or during other situations where they would be at increased risk of **VTE**.

If a person who has **protein C deficiency** has a **VTE**, they will need to have blood thinners. It is important for patients with **protein C deficiency** to have a fast-acting **blood thinner** such as **heparin** started before starting **warfarin** (an oral **blood thinner**). **Warfarin** alone may initially make the patient more likely to clot than less, until the appropriate levels have been reached. Therefore, a **fast acting anticoagulant** is used first and then stopped once the **warfarin** level is adequate.

Protein S Deficiency

Protein S acts with **protein C** to keep the body's natural clotting process controlled. A low **protein S** level has similar effects as a low level of **protein C**.

How common is it?
About 0.2% of the general population has **protein S deficiency**. In patients with **DVT** or **PE**, up to 5% have **protein S deficiency**.

How and when do you test for it?
The level of **protein S** can be tested from a blood sample. However, diagnosis of this condition can be challenging because many things can affect the **protein S** level. Conditions associated with decreased **protein S** include use of **warfarin** (an **oral blood thinner**) and contraceptive use, pregnancy, liver disease, nephrotic syndrome, and severe clot formation.
As with **protein C** and **antithrombin**, a normal test at the time of a new clot rules out the disease, but an abnormal test must be repeated after **warfarin** has been stopped for 2-4 weeks. If it is still abnormal on re-check, then the diagnosis can be made.

What is the risk of VTE in a person with protein S deficiency?
Varying rates of **DVT** or **PE** have been reported with **protein S deficiency**. It is difficult to study given that it is so uncommon. The risk of **VTE** in **protein S deficiency** has been reported from 0 to 11.5 times over that of patients without deficiency. There is no proven increase in artery clot with **protein S deficiency**.

How do you treat a person with protein S deficiency?
As with **protein C deficiency**, patients with **protein S deficiency** who have never had a **VTE** don't need any specific treatment, but if they are to be in any situation that would put them at risk for **VTE**, they should have preventative blood thinners.

A person who has an episode of **VTE** should have a **fast-acting blood thinner** such as **heparin** started prior to starting a longer term **blood thinner** such as **warfarin**.

GROUP 2 DISORDERS:

AN INCREASED AMOUNT OF PRO-CLOTTING FACTORS IN THE BLOOD

Activated Protein C Resistance/Factor V Leiden Mutation

Activated Protein C (APC) resistance refers to the resistance of **Factor V** (one of the proteins in the blood that helps to regulate clot formation) to the activated protein C in the clotting reaction. Since **activated protein C** works on **Factor V** to slow down the clotting reaction, resistance to this causes increased risk of clotting. The majority of **APC resistance** is due to the **Factor V Leiden mutation**, which is a mutation in the gene that codes for Factor V.

How common is it?
Factor V Leiden is the most common inherited **blood clotting disorder**, with an especially high occurrence in people of Caucasian or European descent. It occurs in about 5% of Caucasians, 1.2% of African Americans, 2.2% of Hispanic Americans, 1.2%

of Native Americans, and 0.45% of Asian Americans. In patients with **VTE**, 10-20% have this gene mutation.

How and when do you test for it?
There is a test that looks at whether the **Factor V** is resistant to the **APC**, and there is a genetic test for the **Factor V Leiden mutation**. Either test can diagnose the condition.

What is the risk of VTE with APC resistance?
APC resistance is a relatively weak risk factor for abnormal clot formation. People with one abnormal gene for **Factor V** have a 3 to 7 fold increased risk of clot, and people with both genes abnormal have a 50 to 100 fold increase in risk. The lifetime probability of a symptomatic **DVT** or **PE** in patients with one abnormal gene for **Factor V** is about 10%, thus the vast majority of patients with this condition will never have a **DVT** or **PE**.

However, when a person with **APC resistance** has another **risk factor** for clot, such as oral contraceptive use, hormone replacement therapy or pregnancy, they have increased risk for abnormal clot formation. **APC resistance** may increase the risk of recurrent pregnancy loss and obstetric complications. It is also associated with poorer outcomes in kidney transplant recipients. Whether this condition is associated with abnormal clot in the arteries is unknown.

Prothrombin Defects; Prothrombin Gene 20210A Mutation

The **prothrombin G20210A mutation** is an inherited defect of the gene for **prothrombin**. **Prothrombin** is a protein in the blood that helps clot to form. A person with this condition has high levels of **prothrombin**, which increases the risk of abnormal clot formation.

How common is it?
This disorder is the second most common **inherited clotting disorder**. It is present in 2% of Caucasians, 3% in people of southern European descent, and rare in Native Americans, Asian-Americans or African-Americans. Between 5 to 10% of patients with **VTE** have this disorder.

How and when do you test for it?
There is a blood test that can find the defect in the gene. This test can be accurately performed at any time before, during or after a clot has formed.

What is the risk of VTE with this disorder?
The risk of abnormal clotting is relatively low, with a 2 to 3 times increased risk of **VTE**. Most patients with this **prothrombin gene mutation** will not have had an episode of **VTE** by age 50. Half of clotting episodes in patients with this disorder occur around surgery, trauma, prolonged immobilization, pregnancy or estrogen therapy. This disorder doesn't appear to increased the risk of abnormal clot in an artery.

Factor Elevations (Elevations in the levels of different proteins in the blood that participate in the clotting process.)

The elevation of **coagulation Factors V, VII, VIII, IX, X, and XI** can occur. The association of these with increased risk of clotting is unclear, but persistently high factor levels are more common in patients with a history of VTE.

How and when do you test for elevated factor levels?
There are blood tests available for all of the factor levels. Knowing what the results mean is more difficult. Many things can affect the factor levels. This makes diagnosis of an ongoing elevation of one of these factors challenging. Conditions that can affect factor levels include **Vitamin K** deficiency, malnutrition, liver disease, biliary disease, oral contraceptive use, pregnancy, abnormal cholesterol, obesity, aging, stress, chronic inflammation, recent aerobic exercise, and blood type. Therefore, interpreting the levels of these factors is difficult, as is making the diagnosis of persistent elevated factor levels.

What is the risk of VTE with this condition?
Elevated levels of **Factors V and VII** have not been clearly associated with abnormal vein clot formation, but may be associated with abnormal artery clot formation. In contrast, elevation of **Factors VIII, IX,** and **XI** likely increase the risk of **VTE** slightly. **Factor VIII** elevation has the strongest association with **VTE** with the risk of **VTE** increasing as the level of **Factor VIII** increases.

Hyperhomocysteinemia

Hyperhomocysteinemia refers to an acquired or inherited elevation of the level of the amino acid **homocysteine**. Amino acids are the building blocks that make up proteins in the body. **Homocysteine** is one of several types of amino acids. Acquired **hyperhomocysteinemia** can occur with certain medical conditions, such as kidney failure, hypothyroidism, folate deficiency or Vitamin B6 or B12 deficiency. Inherited hyperhomocysteinemia results from mutations in the genes coding for enzymes that break down homocysteine. These enzymes are **methylene-tetrahydrofolate reductase (MTHFR)**, **cystathione B sytnthase (CBS),** or **methionine synthase**. Defects in these enzymes may or may not lead to **hyperhomocysteinemia**, depending on their severity. **Hyperhomocysteinemia** is associated with both artery and vein clotting problems. How **hyperhomocysteinemia** affects blood clotting is not fully known.

How common is it?
Up to 50% of the general population may have one mutation affecting the metabolism of **homocysteine**. This doesn't result in high **homocysteine** levels in every case.

How and when do you test for this condition?
The diagnosis of **hyperhomocysteinemia** is based on the blood level of **homocysteine**. Levels of **homocysteine** may be elevated for several months after a new **VTE** event, so testing must be done several months after the **VTE** is discovered in order to be accurate.

What is the risk of VTE in a person with hyperhomocysteinemia?
Hyperhomocysteinemia is associated with both abnormal artery and vein clotting, although it is not clear whether **hyperhomocysteinemia** is just a sign of the clotting, or a cause of the clotting. The **homocysteine** level can be lowered with medication, but this doesn't change the risk of **VTE**, therefore the value of testing for this condition is unclear.

How do you treat Hyperhomocysteinemia?
High **homocysteine** levels can be decreased with folic acid, vitamin B6 and vitamin B12 therapy. However, the value of lowering the **homocysteine** level is in question.

Other Inherited Clotting Disorders

There are likely other **inherited clotting disorders** that are not yet fully known. As more of these disorders are figured out, many of the people who now get a **DVT** or **PE** for "unknown" reasons will likely be found to have an inherited condition that increases **VTE** risk.

AntiPhospholipid antibody Syndrome (APS)

Antiphospholipid antibodies are a family of antibodies that are directed against proteins in the blood that are important for coagulation. These antibodies include the lupus anticoagulants and the anticardiolipin antibodies. **Primary APS** includes patients who have **APS** but do not have lupus or other autoimmune diseases. **Secondary APS** includes those patients with **APS** and systemic lupus erythematosus (SLE).

How common is it?
This condition is reported in only 2% of healthy individuals, but is found in up to 20% of patients with **VTE**.

How and when do you test for this?
Diagnosis of this condition is based on both clinical and laboratory tests. In order to be diagnosed with this condition a person must have elevated antibody levels on two tests at least 6 weeks apart, and they must have had an abnormal clotting event or have had pregnancy related complications.

What is the risk of VTE in people with APS?
The risk of **VTE** in people with **APS** is relatively high. Approximately 1/3 of people with **APS** have had an abnormal clotting event. Usually this event is a **DVT**. People with **APS** who do not have lupus have an 11-fold increase in risk of **VTE** over those who do not have **APS**. They also have an increased risk of artery clot formation. Patients with lupus are at increased risk of abnormal clot formation even if they don't have the antibodies.

How do you treat APS?
People with **APS** who have a first **VTE** are usually treated with a short acting **blood thinner** such as **heparin**, and then change over to **warfarin** therapy after 5 days. The length of time to continue the **blood thinners** after the first abnormal clotting event is somewhat controversial and can range from 12 months to lifelong. In patients with **APS** who have not had a **VTE** event, daily aspirin therapy is recommended, but if other additional risk factors exist, **warfarin** or **heparin** therapy should be considered.

Heparin Induced Thrombocytopenia

Heparin induced thrombocytopenia (HIT) is a severe side effect of **heparin** therapy that can cause abnormal clotting. This condition occurs when a person's body makes an antibody against the **heparin**, and that antibody also targets their platelets. The antibody binding to the platelets causes them to clump up, which forms a clot.

How common is HIT?
Although **heparin-induced antibodies** form in 10-20% of people who get heparin, most of these patients do not develop **HIT**. Only 1-3% of people who use **heparin** for 5 days get **HIT**. This can go up to 6% after 14 days of continuous use. **Low-molecular weight heparin** (a more purified form of **heparin**) has a lower risk of **HIT**, but is harder to reverse and is more expensive.

How and when do you test for HIT?
A person who develops a new clot while on **heparin** is suspicious for **HIT**. A falling platelet count in the blood is another sign of **HIT**. In **HIT**, platelet counts fall starting 5-10 days after the start of **heparin** therapy, and reach a low by 7-14 days. This is known as "typical-onset" **HIT**. "Delayed-onset" **HIT** occurs when the platelet count falls later (up to 20 days after **heparin** starts) and can even occur after a patient has stopped the **heparin**. "Rapid-onset" **HIT** can occur within 24 hours of starting **heparin**. This can occur in patients who have had **heparin** before. A drop in the number of platelets by 50%, or a fall to below 100,000 is considered suspicious for **HIT**. To make the diagnosis of **HIT**, a lab test needs to be done. There are several available tests, some that look at platelet function, and one that looks for the antibodies themselves. If a patient is suspected of having **HIT**, the heparin should be stopped immediately while testing is done.

What is the risk of VTE in a person with HIT?
The most common complication of **HIT** is abnormal clot formation. About 50% of patients with **HIT** will develop a clot or die within 30 days if not treated. The risk of abnormal clot formation is increased 30 times in **HIT** patients. The most common clotting event is **DVT**. **PE** is also common in these people. Abnormal artery clotting is less common. Abnormal skin lesions can form in patients with **HIT** about 20% of the time.

How do you treat HIT?
The first thing to do is to stop all sources of **heparin**. Also, a different **blood thinner** should be started to help prevent the abnormal clot formation. Other **blood thinners** that could be used include **lepirudin, argatroban** and **bivalirudin**. **Low-molecular weight heparin** is still a **heparin**, and should not be used. Once the platelet count has come back up to normal, **warfarin** can be started, but should overlap with the other blood thinner by 5 days.

A person who has had **HIT** should not get heparin again in their life, unless under rare and very special circumstances.

Cancer

VTE is a major cause of complications in **cancer** patients. **PE** is the cause of death in one of seven hospitalized **cancer** patients who dies. The risk of **VTE** is much higher in **cancer** patients than in non-**cancer** patients, and most of the clots occur without another **risk factor** being present. Surgery, chemotherapy, central venous line placement, and immobility all further increase the risk of clotting in **cancer** patients. Treatment of **VTE** in cancer patients should continue until the **cancer** is in remission and no further chemotherapy is planned.

CONCLUSION

There are many different factors that can increase the risk of **VTE**. Some of these increase the risk more than others. These conditions should be looked for in any person who has a **VTE**, unless the cause is already known.

CHAPTER 4
DEEP VEIN THROMBOSIS PREVENTION

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Introduction

Deep vein thrombosis (DVT) and **pulmonary embolus (PE)** are serious conditions. **Venous thromboembolism (VTE)**, which is either **DVT** or **PE**, is the 4th leading cause of death in Western society. People who get a **DVT** have a 30% chance of getting a second **DVT** within 10 years. Thus, preventing **VTE** is very important. This chapter will discuss the main ways to prevent a **DVT** or **PE**.

Risk Factors

There are many factors that can increase one's risk for **VTE**. The biggest risk factors include hospitalization, surgery, cancer and **hypercoagulable disorders** (factors in your blood that increasing your chance of developing blood clots).

Prevention of DVT and PE

There are two types of **DVT** and **PE** prevention. The first is called "**mechanical**" and the second is called "**pharmacologic**", which means through the use of medication. **Mechanical prevention** includes the use of **compression stockings**, leg elevation, **sequential compression devices (SCD's)**, ambulation, and **vena cava filters**. **Compression stockings** are tight elastic stockings that are worn by people in the hospital or by people who can't be active to prevent a **blood clot**. They work by decreasing the "pooling" of blood in the legs. Leg elevation and **SCD's** work the same way. **SCD's** are plastic devices that are fit on the legs, and they inflate in a sequential way (i.e. from bottom to top) in order to empty the **veins** in the legs. These methods have been shown to decrease the number of **DVT**'s in hospitalized patients by 20%.

Vena cava filters are another form of **mechanical prevention**. The vena cava is the large vein in the abdomen that brings blood back to the heart and lungs. A **vena cava filter** is a metal device that resembles an umbrella. This can be placed in the vena cava to catch any clots that might break off from the legs, and prevent it from going to the heart. This filter can't prevent **DVT**, but it can prevent **PE**. This device is generally used in patients who have a **DVT**, or who are at high risk of **DVT**, but can't get **blood thinners** for one reason or another.

Pharmacologic prevention basically includes **blood thinners**. The most common type of **blood thinner** used to prevent **VTE** is **heparin**. There are 2 forms of heparin: **unfractionated heparin**, which is given IV, and **fractionated heparin** (also known as **low molecular weight heparin, LMWH**) which is given by an injection under the skin. Another type of **blood thinner** given by injection is called **Fondaparinux**. This is a

synthetic heparin-like drug. There are also a few other medications that can be given to prevent clot in those people who are allergic to heparin. These are called **lepirudin**, **bivalirudin** and **argatroban**. Which of these medications is used for clot prevention depends on the patient and the situation.

Another commonly known blood thinner is **Coumadin** (also known as **Warfarin**). **Coumadin** is not generally used to prevent **VTE** in a patient who has not had **VTE** in the past, but it is used to prevent recurrent **VTE** in a patient who has already had one.

Aspirin, Plavix, or other drugs that affect platelet function, are not used to prevent **VTE**. They are not as effective as the other medications already mentioned.

Who should get preventative treatment?

There are very well established recommendations for who should get **VTE** prevention. For patients who are having surgery, the recommendation for preventative treatment depends on the type of surgery, the age of the patient, and whether or not the patient has risk factors for clot formation. This risk stratification is described below.

Low Risk Patients

Patients who are having low risk surgery (usually surgery that lasts less than 30 minutes, or is not extensive), who are under 40, and who have no other risk factors are considered low risk for developing **VTE**. These patients generally don't need medication for **VTE** prevention. They are encouraged to walk early after surgery.

Moderate Risk Patients

Patients who are having major surgery, who are older than 40, but have no additional risk factors are considered moderate risk. These patients are also encouraged to walk early if possible after surgery, but they are also given low dose **blood thinners** (usually either **unfractionated heparin** or **LMWH**) around the time of surgery.

High Risk Patients

Patients who are having major surgery, who are older than 40, or have additional risk factors are considered high risk. These patients are encouraged to walk early if possible, to wear compression stockings, and to get low dose **blood thinners** around the time of surgery.

Very High Risk Patients

Patients older than 40, who are having major surgery, and have major additional risk factors (examples: cancer history, paraplegia, hip or knee surgery, known clotting disorder), are considered very high risk. Not only should these patients be given **blood thinners** around the time of surgery, they should also have **SCD's** on, and can also be asked to wear **compression stockings**.

Non Surgical High Risk Patients

Hospitalized patients who are not having surgery are also at risk for **VTE**. The highest risk is in those patients with severe heart failure, chronic obstructive lung disease, severe infection, cancer or paralysis. Risk also increases with age, and is higher in patients who have had a **VTE** episode in the past.

Are there special circumstances that change the risk of DVT?

Hip or Knee Replacement Surgery

Patients undergoing hip replacement surgery or knee replacement surgery are at especially high risk for **DVT** and **PE**. Often these patients are given higher doses of **blood thinners** around the time of their surgery to help prevent clot formation. This must be balanced with the risk of **bleeding** if too much **blood thinner** is used. However, the vast majority of these patients should have **blood thinners** around the time of surgery.

Neurosurgery

Any **bleeding** after brain surgery or back surgery could be very dangerous, therefore **blood thinners** are not generally used in most of these patients Because the consequences are so high if there is any **bleeding**, **SCD's** with or without **compression stockings** are used for clot prevention. In those patients who are at high risk for clot formation, low doses of **blood thinners** may be used around the time of surgery.

Trauma

Patients who have had severe trauma are at high risk for clot formation. Many of these patients cannot get **blood thinners** because of the risk of **bleeding**. In those patients, a **vena cava filter** may be used along with compression stockings and **SCD's**.

How long preventative measures should be given?

In general, the preventative measures, whether it be mechanical or medications, should be continued until the risk is over or significantly decreased. This is generally thought to be once a hospitalized patient is walking several times per day. In a patient with cancer, the prevention should be continued until the cancer is in remission.

Conclusion

DVT and **PE** are one of the most preventable causes of death and disability. Prevention should be used appropriately, and if it is, **DVT** and **PE** can be prevented in most patients.

CHAPTER 6
CLINICAL PRESENTATION OF VENOUS THROMBOSIS "CLOTS": DEEP VENOUS THROMBOSIS AND PULMONARY EMBOLUS

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Abstracted by Kellie R. Brown

Introduction

The body has normal processes that balance between **clot** formation and clot breakdown. This allows **clot** to form when necessary to stop bleeding, but allows the **clot** formation to be limited to the injured area. Unbalancing these systems can lead to abnormal **clot** formation. When this happens **clot** can form in the deep veins usually, but not always, in the legs, forming a **deep vein thrombosis (DVT)**. In some cases, this **clot** can dislodge from the vein in which it was formed and travel through the bloodstream into the lungs, where it gets stuck as the size of the vessels get too small to allow the **clot** to go any further. This is called a **pulmonary embolus (PE)**. This limits the amount of blood that can get oxygen from the lungs, which then limits the amount of oxygen that can be delivered to the rest of the body. How severe the **PE** is for the patient has to do with the size of the **clot** that gets to the lungs. Small **clots** can cause no symptoms at all. Very large clots can cause death very quickly. This chapter will describe the symptoms that are caused by **DVT** and **PE**, and discuss the means by which these conditions are diagnosed.

What are the most common signs and symptoms of a DVT?

The symptoms that are caused by **DVT** depend on the location and extent of the **clot**. If the **clot** is small, or if it is limited to the small **veins** in the calf, there may be no symptoms at all. If the **clot** is extensive involving the thigh **veins** and/or the large **veins** in the pelvis the symptoms can be very extreme.

The most common **symptoms** a person experiences when they have a **DVT** are pain and **swelling** in the involved extremity. This can be subtle ankle and calf **swelling** with minimal pain, but if the **clot** is extensive the entire leg can be very swollen, tight, and painful.

Other **symptoms of DVT** include redness, tenderness, unexplained fever, increased visibility of skin veins, or bluish discoloration. Pain in the calf when the toes and foot are stretched upward is another sign of **DVT**. This is called a **Homan's sign**, and it is not reliable in diagnosing a **DVT**.

Unfortunately, diagnosing **DVT** by **clinical signs** and **symptoms** is notoriously inaccurate. The **symptoms** caused by **DVT** are vague and non-specific and up to 50% of patients with **DVT** have no symptoms at all. Therefore, a low threshold to get further testing is appropriate if there is a suspicion of **DVT**.

What are the most common signs and symptoms of a PE?

One of the most feared complications of **DVT** is **pulmonary embolus**. **PE** occurs in about 10-25% **DVT**'s. Although sometimes the only symptoms of **DVT** experienced by a patient are those of a **PE**, most **PE**s may be asymptomatic. The **symptoms of PE** include a sudden onset of chest pain, shortness of breath (breathing very fast) and increased heart rate. Sometimes a person with a **PE** will pass out from the **PE**. Other less common signs are pain with breathing, dizziness and anxiety. Most of these **symptoms** are very vague, and could be due to a number of different conditions. Therefore, other tests are needed to find a **PE**. A person who experiences a sudden onset of these symptoms should be evaluated immediately.

How is a DVT diagnosed?

If after doing a history and physical exam there is a suspicion of **DVT**, further testing is indicated. The most common test to diagnose a **DVT** is an **ultrasound** (sound wave study) of the legs, abdomen or arms. What the doctor is looking for are parts of the **veins** that appear swollen and do not compress with pressure and show abnormal blood flow. The test does not require a stick into the body and is consider non-invasive since it only requires an **ultrasound** device placed on your skin. **CT scan (computerized tomography)** or **MRI (magnetic resonance imaging)** can also be used to make the diagnosis but are more costly and the **CT** requires a special agent into the body to see the veins. In the past, the lower leg **veins** were actually punctured and special **X-ray** drug was placed into the **vein** to see the insides of the **veins** but this is rare done in current medical practice.

How is a PE diagnosed?

The first tests that are usually done in people who have symptoms of a **pulmonary embolus** are a **blood gas test** (this tests for the amount of oxygen in the blood), and an **EKG** (this tests for a heart attack). These tests are usually very fast, and can help the doctor decide if the **symptoms** are from a heart attack, or from a **pulmonary embolus**. Another blood test, called the **d-dimer test**, can also be done. The **d-dimer test** is really helpful if it is negative. This means that a **DVT** or **PE** is really unlikely. However, if it is positive, that doesn't mean that a **DVT** or **PE** is present as other conditions can produce an elevated **d-dimer** result. Also, the result of the **d-dimer test** is not back immediately, so sometimes other tests are done before this result is back, and if the **d-dimer** is positive, then other tests are definitely done.

The next step is to get a specific finding of a **pulmonary embolus**. This is usually done by getting a **CT scan** immediately after injecting a drug into a **vein** which helps the doctor to see the inside of the lung blood vessels. If the doctor sees abnormal filling of the lung blood vessels than a **clot** is present. **CT scan** is very good at finding a **PE**, and can also be used to look for a **DVT** in the upper legs. If **CT** cannot be done due to an allergy to the contrast dye, an **MRI (magnetic resonance imaging)** study can be done to test for **PE**. The **MRI** uses the magnetic properties of blood to see the inside of the lung

veins. Another possible test to look for **PE** is called a **ventilation-perfusion scan** (it is also called a **VQ scan**). This test uses a special drug to see the lung blood vessels with a radiation scanner. This test is not as good as a **CT** scan, but may be done in people who are allergic to the contrast dye, or in pregnant women.

A **pulmonary angiogram** is the most accurate study for **PE**, but it is also the most risky. This test involves putting a catheter into a blood vessel in the groin, and passing it up to the heart and injecting dye into the blood to see the arteries of the lungs. Because this test is risky, it is usually only done in situations where the catheter is used to try to get the **clot** out of the lungs. This is only done in the most severe cases.

Conclusion

The most common **symptoms of DVT** are pain and **swelling**, but many **DVT**s have no **symptoms**. The most common **symptoms of PE** are sudden onset of chest pain and shortness of breath. A low threshold to get further tests is needed in order to diagnose most **DVT**s because the symptoms are vague. DVT is usually diagnosed with ultrasound, and PE is usually diagnosed with CT scan, but other tests may be needed to make the diagnosis.

CHAPTER 5
SUPERFICIAL VENOUS THROMBOSIS

Original authors: Anil P. Hingorani and Enrico Ascher
Abstracted by Gary W. Lemon

Clinical Presentation /Diagnosis

Superficial venous thrombophlebitis (SVT) is an attack of **blood clots** in **veins** that lie under the skin within the fat of the body but not in the **deep veins**. It is a relatively common medical problem. It can get better and then happen again (recurrence). It has a low chance but known risk of **deep vein thrombosis** (**DVT**, **blood clots** in the **deep veins**) and **pulmonary embolism** (**blood clots** moving to the lungs). How often **SVT** occurs in the population is unknown. It affects men and women at about the same rates with most occurring in the mid-fifties. **Varicose veins** are present in over sixty-two percent of patients having **SVT**. Other risk factors (factors that increase the risk of having **SVT**) include greater than 60 years old, obesity, smoking, and a prior history of **DVT** or **SVT**. The physical signs of **SVT** is warmth and redness with pain along a **superficial vein** and sometime even a cord or thickening of the vein that can be felt because the blood clot within is making the vein bigger in size. Swelling of the lower leg or arm may or may not be present. Other disease conditions such as infection of the skin and fat (**cellulitis**) or infection of the third type of blood vessel (**lymphangitis**) may look like **SVT**. However, a **duplex ultrasound** (a sound wave study used to see the **vein** though the skin) will help to know if **venous thrombosis** is present. So the diagnosis is made by the presence of associated symptoms, physical findings, and showing the blood clot within the vein by **duplex ultrasound**.

The treatment of **SVT** depends on the location of the **venous thrombosis** and what caused it. The most common cause of **SVT** in the upper extremities is from an intravenous catheter (plastic tube placed into the **vein**) used to give the patient fluids or medicines. **SVT** can also occur in the large **veins** alongside the breast and chest wall. It can also develop at various sites in the lower leg. It is not uncommon for these events to be associated with an underlying cancer or **hypercoagulable** (increased blood clotting) **states**. **SVT** of the lower extremities can be found in branches of the **greater saphenous vein** (the large vein going from groin to ankle which lies in the fat just under the skin) only, involving the **greater saphenous vein** and its offshoot vein. It can also happen in the **small saphenous vein** located in the back of the calf. **SVT** can occur in **varicose veins** with or without an **ulcer** present in the lower leg.

Once physical findings suggest that **SVT** is present, this clinical idea is proven by a venous **duplex ultrasound** study. A **duplex ultrasound** study uses sound waves to make a picture of the vein and to show if blood is flowing in the vein or not. No blood movement and a large vein show that **SVT** is present. A **venogram** involves placing a needle in a vein further out from the suspected clot and injecting agents which can be seen by X-ray to show where the clot is located. This is rarely used in current medical

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practice. **Duplex ultrasound** has taken it place, is safe, can be repeated as needed, is completely noninvasive (does not invade the body) and is very accurate in showing the **superficial vein thrombosis**. **Duplex ultrasound** should be done whenever **SVT** is found in the legs. Up to forty percent of patients with leg **SVT** may also have a **deep vein thrombosis** which is more risky for the patient.

Etiology

The cause of **SVT** is not always easy to find. Certainly injury to a **varicose vein** from trauma (hitting) or the presence of an IV catheter in an area where **SVT** is found is the likely cause. But, in about forty percent of patients there is a **hypercoagulable state** which is causing the increase risk of **blood clots**. Identifying which abnormal **clotting factor** is present may change how the doctor must treat the **SVT**.

Treatment

The location of the **SVT** will help the doctor decide on what local care or drug and how long the treatment should be given. **SVT** caused by an intravenous catheter can be treated by taking the catheter or needle out of the vein and placing warm cloths (compresses) over the area. Non-steroidal anti-inflammatory drugs, such as ibuprofen, taken by mouth for a short time helps to relieve the discomfort and decrease the local affects. Antibiotics may be necessary if pus is noted from the drainage site around the IV catheter when removed. Occasionally infection which can not be cleared from the vein with these methods and results in making the person very sick (sepsis) may require taking the vein out with an operation. This is rarely needed.

Recurrent **SVT** occurring in varying sites of the lower extremity or the presence of **SVT** along the chest wall or breast area is most commonly treated with warm compresses and non-steroidal drugs. Work up for these conditions should include a search for an unknown cancer or state which is causing the patient to be too active in forming blood clots (hypercoagulable state).

There are still questions as to how best to treat **SVT** involving the lower extremities which appear to have no known cause and in some particular locations within the body. **SVT** of the lower extremities not associated with **deep vein thrombosis**, having no **hypercoagulable state** and occurring in a place not near the groin or upper thigh are most often managed with warm compresses, compressive stockings, and non-steroidal drugs. **SVT** with a **deep vein thrombosis** and/or **hypercoagulable conditions** and/or **venous thrombosis** in the **saphenous vein** near the groin and upper thigh or upper calf are given full dose **blood thinning drugs** (**anticoagulants** such as **heparin**). A **duplex ultrasound** is obtained at the first thought that **SVT** is present to make sure these conditions are not present and if **anticoagulant** drugs are not given it is repeated in 48 to 72 hours to make sure that the clot has not gone up the **vein** or deeper in the body. **Anticoagulant** therapy with **heparin** and long tern **blood thinning** with **warfarin** may be used if the clot is extending. Occasionally, surgical removal of the clotted **great saphenous vein** with tying it off in the high groin may be suggested as another way to

decrease pain and stop any more clotting from taking place. This may become necessary if the patient cannot take anticoagulant medication for medical reasons.

Conclusion

Most cases of **SVT** are not noticed or taken seriously by the patient and get better without treatment. It found, the treatment is often conservative with the use of **compression stockings**, non-steroidal drugs, and removing an IV catheter if this is the cause. It is important to make sure that the patient does not have a condition which is causing more than normal blood clotting (**hypercoagulable states**) or has an unknown cancer. Finally, **anticoagulant** medication may be necessary for those **SVT's** that are in the largest part of the **saphenous vein** (near the groin or upper thigh), are increasing up the **vein** during observation or those who are also found to have a **deep vein thrombosis** or **pulmonary embolism**. If there is no treatment with a **blood thinning drug**, a repeat **duplex ultrasound** study should be obtained in most patients with lower extremity SVT.

Commonly asked questions

What is superficial venous thrombosis?

Superficial venous thrombosis is a **blood clot** forming in one or more **veins** located in the fatty tissue lying directly below the skin. These are usually rather small **veins** and the clot stops within these **veins**. If the blood clotting stops before it gets to the largest part of the major **superficial vein** (the **saphenous vein**) or before it goes to the **deep veins**, it generally will not cause much more than local pain, swelling, and redness over the **vein**.

How do I know if I have superficial venous thrombosis?

You will usually have a tender area over a **vein** and may see local redness with some swelling of the lower leg. When you go to the doctor to have it further studied, the doctor will usually obtain a **duplex ultrasound** study which uses sound waves to have a picture of your **veins** and to show if blood is moving normally in the **veins**. Venous clotting (**thrombosis**) is present when the **veins** are bigger than normal (filled with **blood clot**) and little or no blood movement is taking place.

What is the treatment of superficial venous thrombosis?

If the **SVT** is located only in a lower leg **varicose vein** and not close to the big part of the biggest **superficial vein** (the **saphenous vein**), treatment is usually warm compresses, **compressive stockings** and antiflammatory drugs to decrease local discomfort. If **SVT** goes into the biggest **superficial veins** especially close to the groin and upper thigh or if there is also **deep venous thrombosis**, than full dose **blood thinning agents** (**anticoagulant** therapy) are needed to protect you from blood clot going to the lungs (**pulmonary embolus**).

CHAPTER 7
MEDICAL TREATMENT OF DEEP VEIN THROMBOSIS AND PULMONARY EMBOLUS

Original authors: Russell D. Hull, Graham F. Pineo, and Thomas W. Wakefield

Abstracted by Kellie R. Brown

Introduction

Once the diagnosis of **deep vein thrombosis (DVT)** or **pulmonary embolus (PE)** is made, treatment of the condition must be undertaken. While new therapies are on the horizon, this chapter will discuss currently accepted therapies.

Goals of Treatment (Why treat a DVT or PE)?

- To prevent a **pulmonary embolism** (if not already present)
- To prevent death from **pulmonary embolism**
- To prevent a recurrent **DVT**
- To prevent the **post-phlebitic syndrome**

How is a DVT or PE treated? (See Table 1)

DVT and **PE** are treated essentially the same, with a few exceptions. Once the condition is diagnosed, **heparin** (a **blood thinner** injected into a **vein** or directly into the fatty tissues of the body) is given. There are two types of **heparin** that can be used. One is called **unfractionated heparin**, and this is the one that is runs directly into the **vein** by way of an intravenous line. One is called **Low Molecular Weight Heparin (LMWH)**, and this is the one that is injected directly through the skin into the fatty tissues of the body. Either can be used, but **LMWH** is generally preferred, unless the patient has kidney failure.

Once the **heparin** has thinned the blood enough (as determined by blood tests), an oral **blood thinner** can be started. This is called **warfarin (commonly known as Coumadin)**. The **heparin** is continued until the **warfarin** has reached an effective level, as determined by a blood test called the **INR** (which stands for **international normalized ratio**). The **INR** should be about 2 to 3 in most cases. At this point the **heparin** can be stopped.

The **warfarin** is continued for various amounts of time, depending on the clinical situation. In most uncomplicated cases of a first time **DVT** or **PE**, the **warfarin** is continued for 3-6 months. In certain cases, when the **DVT** or **PE** is recurrent, or when the risk factors for **DVT** or **PE** are not temporary (such as a blood clotting disorder), **warfarin** may be maintained for life.

What if blood thinners can't be used?

If **heparin** or **warfarin** can't be used, either because the patient can't tolerate it or they have a high risk of bleeding from it, then a **vena cava filter** can be used. **A vena cava filter** is a mesh "umbrella" device that is inserted into the large **vein** in the abdomen using a catheter inserted in the groin. This catches any **clot** that may break off from the **clot** in the leg, and prevents it from traveling to the lung and becoming a **PE**.

Surgery to remove the **clot**, either from the leg (as in a **DVT**) or from the lung (as in a **PE**) is rarely done. If the symptoms from the **DVT** or **PE** are very severe and life or limb threatening, then **thrombolysis** (**clot** dissolving drugs) can be considered.

Some patients with a **pulmonary embolus** are very ill. If a patient with a **PE** has a very low blood pressure, or is having a lot of trouble keeping enough oxygen in the blood, then a more aggressive treatment may be done. One option for more aggressive treatment is an injection of a medication to break up clot. This is called **thrombolysis**. This can break up the clot in the lung arteries, but it could also break up clot in other places in the body, so it can cause bleeding. The most dreaded complication of this treatment is to bleed into the brain. Because of the risk of this, **thrombolysis** is used only if the patient is not able to keep up their blood pressure or oxygen level.

Another aggressive treatment that could be tried is to remove the clot from the artery that has the **PE** in it. This can be done using a catheter placed from the groin and passed up into the lungs, or it can be done with surgery. This is only done in patients who are likely to die without this treatment.

Is there anything else that can be done to help the symptoms?

In addition to medication or a **vena cava filter**, patients who have a **DVT** should wear **elastic compression stockings** for 2 years after the DVT is diagnosed. This will help prevent the **post-thrombotic syndrome**; long-term **swelling**, lower leg **skin changes**, or even skin breakdown (**ulcers**). Also, patients who have a **DVT** can walk around as usual. Bed rest is not encouraged. Leg elevation when at rest is helpful.

What if the DVT is in the arm?

Most **DVT**s are located in the legs. However, **DVT** can occur in the arm. If it does, it should be treated just as lower extremity **DVT**'s are treated. Most **DVT**s in the arm are caused by placement of central venous catheters (which are large IVs that are placed in the large **veins** of the arm and neck.) If one of these catheters is present in a **vein** that has **clot** in it, it should be removed and blood thinners should be started just as in the case of a leg **DVT**.
If the **clot** is thought to be due to **thoracic outlet syndrome** (a condition where the opening from the chest into the arm isn't large enough, and the vein gets compressed with arm movement), then **thrombolysis** is often used, followed usually by surgery to make the **thoracic outlet** larger.

Conclusion

DVT and **PE** are treated essentially the same. Initial treatment usually consists of **LMWH**, followed by **warfarin**. Once the **warfarin** levels are appropriate, the **LMWH** is stopped. The **warfarin** is continued for 3-6 months in most cases of a first time **DVT** or **PE**. In the case of a recurrent event, **warfarin** may be continued for life. If **blood thinners** aren't able to be used, a **filter** can be placed to prevent the **clot** from travelling to the lungs. If symptoms are very severe, removal of the **clot** from the **vein**, usually by **thrombolysis**, can be undertaken.

Table 1: DVT and PE Treatment Summary

Initial Treatment of DVT or PE

- Low molecular weight heparin (LMWH) or IV heparin should be started.
- This should be continued for at least 5 days.
- Warfarin (an oral blood thinner) can be started once the heparin is at appropriate levels.
- Heparin can be stopped once the Warfarin is at an appropriate level (INR of 2-3).

Additional Treatments

- Thrombolysis is not routinely used.
- Thrombolysis can be considered if the clot is severely symptomatic, or if it life or limb threatening.
- Vena Cava Filter can be used if blood thinners cannot be used, or if they fail.
- Surgery to remove the clot is not routinely used. It may be considered in very severe cases.

Length of Warfarin Therapy

- For patients with a first time DVT or PE due to a reversible risk factor, treatment with Warfarin is recommended for at least 3 months.
- For patients with a first time DVT or PE who have no identifiable risk factors, treatment should continue for at least 6 to 12 months.
- Patients with a DVT or PE who have no identifiable risk factors should consider being tested for a clotting disorder.
- For patients with a first time DVT or PE who have a known clotting disorder should be treated for at least 12 months, and possibly indefinitely.
- For patients with more than one known DVT or PE episode, warfarin therapy should be continued indefinitely.

Post-thrombotic Syndrome

- An elastic compression stocking with a pressure of 30-40mmHg at the ankle (high grade compression) should be used for 2 years after an episode of DVT. This helps to prevent long-term symptoms.

CHAPTER 8
SURGICAL/INTERVENTIONAL TREATMENT OF ACUTE DEEP VENOUS THROMBOSIS

Original authors: Anthony Comerota, Bo Eklof, Jorge L. Martinez, and Robert B. McLafferty

Abstracted by Michael C. Dalsing

Introduction

Acute deep venous thrombosis (DVT) is the formation of a **blood clot** in one or more of the **deep veins** of the leg and abdomen. The **calf veins** are the most common site of a thrombus (**clot**), but 40% of **DVT**s occur in the **femoral** (thigh) and **iliac** (hip) **veins** starting behind **valves** (which prevent a **reflux** or backflow of blood). Extensive **deep venous thrombosis** causes acute pain, swelling, and leg discoloration and may result in the **post-thrombotic syndrome** (chronic pain, swelling, skin discoloration, and, potentially, skin breakdown generally around ankle (ulcers). The more extensive the **deep venous thrombosis**, the more severe the **post-thrombotic syndrome** will be as one gets older. The **post-thrombotic-syndrome** is the result of both damaged **valves** and blockage to blood flow. Over time, patients with extensive **venous thrombosis** are likely to develop progressive **valvular incompetence**. This is the inability of the vein valves to keep the blood from flowing backward (**reflux**) into the legs after it has started its journey back to the heart. When a **valve** isn't doing its job in preventing **reflux**, it is called incompetent. Removing the **blood clot** is likely to preserve function of the **vein valves** and prevent later **reflux**. Removing the **blood clot** also removes the blockage to blood flow through the **veins** both at the time of the acute **clot** and to prevent the symptoms of the **post-thrombotic syndrome**. The earlier the treatment is begun after a patient develops a **clot**, the more likely the treatment will be successful.

Why Remove Acute Blood Clot? Goals of Treatment

- To prevent **pulmonary embolism** (blood clot in the lung, which can be fatal)
- To decrease pain and swelling of the affected leg
- To prevent or stop the development of **phlegmasia cerulea dolens** and **gangrene** (tissue death and even loss of the leg) from loss of blood supply cause by a total blockage of blood flow through the leg **veins**. **Phlegmasia cerulea dolens**, sometimes called **blue phlebitis**, is an **acute thrombosis** with **edema** (accumulation of fluid), **cyanosis** (blue-discolored skin), and **petechiae** (reddish or purplish spots)
- To prevent the disabling **post-thrombotic syndrome** by removing the **blood clot** (preventing blockage of blood flow) and preserving normal function of venous **valves**.

Ways (Method) of Removing Acute Blood Clot

The first choice for an early and quick removal of a **blood clot** in a **deep vein** is catheter-directed **thrombolysis**. The treatment is designed to break down (dissolve) the blood

clots by injecting a drug into a **vein** or directly into the **blood clot**. The drug used is called **plasminogen activator** and it acts by breaking down (splitting) the body's own **plasminogen** (trapped within the **clot**) into its active form called **plasmin**. **Plasmin** is an enzyme and acts breaking down the **fibrin** (the solid substance of the **blood clot**) so that it can again flow in the blood and be removed by the body. The **plasminogen activator** can be injected into any **vein** in the body by a standard intravenous needle (IV) to be delivered to all parts the body equally (**systemic thrombolysis**). But is now more commonly delivered right into **blood clot** itself by way of a needle puncture of a **vein** through which a long catheter (plastic tubing) can be pushed into the **blood clot**. The **plasminogen activator** can then be pushed (injected) right into the **clot** (**catheter-directed thrombolysis**) for fasted action. One of the risks of this method is that the **plasminogen activator** can breakdown any **clot**, any where in the body so breakdown of a **blood clot** in an area where it might cause trouble (brain, stomach, or following recent eye surgery or other major surgery) can occur. **Percutaneous mechanical thrombectomy** provides a way to improve **catheter-directed thrombolysis** by adding a mechanical mixing or stirring of the **clot** and the **plasminogen activator**. This mixing makes the breakdown of **fibrin** go faster. The devices are delivered into the **clot** in the same way the catheter is pushed into the **clot**. One device works with or without **thrombolysis** (**clot** breaking drug) using simple stirring to break up the **clot** into small pieces and to then suck the **clot** from the body.

When **thrombolysis** fails or is contraindicated (most often because of an increased risk of bleeding), **open venous thrombectomy** is a good alternative. **Open venous thrombectomy** is a surgical removal (pulling out) of a **thrombus** lodged in a **vein** by making a cut through the skin to get to a **vein** through which can be placed a catheter with a balloon on the end. The balloon tipped catheter is pushed past the blood clot; the balloon is filled with fluid to the size of the **vein** and pulled back to pull the clot from the **vein**. The hole in the **vein** used to place the catheter and remove the **clot** is closed with suture (needle and thread). Often, a small connection between a nearby artery and vein (**arteriovenous fistula**) is made to help the **vein** to stay open.

Heparin (a **blood thinner** given by **vein** or by injecting a fat part of the body) is given for several days after surgery. **Warfarin**, a **blood thinner** taken by mouth is started on the first postoperative day and continued for months. The patient walks with a **compression stockings** the day after surgery and usually leaves the hospital in a few days.

Venous narrowing (stenosis) may be seen as one cause of the blood clot

Many patients with **acute deep venous thrombosis** also have an **iliac (pelvic) vein stenosis (narrowing)**. **Thrombolysis** or surgery uncovers the vein narrowing, which can then be corrected in order to keep the vein open and to avoid repeated episodes of **deep venous thrombosis**. The **stenosis** is popped open by placing a catheter with balloon end in the narrowed vein segment and inflating the balloon with fluid, thus performing a **balloon angioplasty**. A **stent** (a slender, metallic mesh cylinder) is placed within the narrowed segment of the **vein** and dilated also. The **stent** keeps the **vein** open and prevents it from collapsing.

Risks connected to removing acute blood clot

The complication of greatest concern when using **thrombolysis** is **bleeding**. Most commonly this occurs from the puncture made in the skin to place the catheter in the **vein**. Other areas of the body, such as the stomach, kidney bladder, or muscles that may have been unexpectedly hit may be areas of **bleeding**. The most feared **bleeding** is intracranial (within the brain) leading to a stroke. Fortunately, this is rare when patients are chosen properly but can still occur. **Pulmonary embolism** is also a rare complication of **thrombolysis** since the **clot** is being broken up into pieces. These risks are also noted when **mechanical thrombectomy** devices are used. The whipping of the blood may break the blood down into its parts which can cause damage to the kidneys (kidney failure) or rarely to other parts of the body. If the open surgery is needed, there is a cut into the body which can bleed (called a **hematoma** if the blood is trapped under the skin), become infected or drain fluids. No matter the method of acute **clot** removal, the **vein** can reclot which is why **blood thinner** are used.

Expected Results

According to recently published reports, patients treated early in their course of **iliofemoral deep vein thrombosis** with **catheter-directed thrombolysis** can expect an 80% success rate compared with 18% success in patients treated with conventional **anticoagulation** (agents taken to thin the blood).

Surgery is often used only in the very symptomatic or after other methods have failed so the expected results may not be as impressive. Studies have shown that about 40% of patients who had **open thrombectomy** and normal working **veins** compared with about 10% of patients treated with **blood thinners** only. After 10 years, **reflux** of blood in the **popliteal vein** (located behind the knee and a bad event) was found in about 30% of patients who had surgery and in about 70% of the group treated only with **blood thinners**.

Conclusion

Generally, **catheter-directed thrombolysis** with or without mechanical device use is the preferred treatment option for patients with **iliofemoral deep venous thrombosis** who are otherwise healthy and have no contraindication to receiving a **thrombolytic drug**. If **thrombolysis** is too high a risk, **venous thrombectomy** is recommended. For patients who are bedridden and those who are in very poor health, treatment with **anticoagulation** agents (**blood-thinning** agents) alone may be advisable. Successful and timely **clot** removal in patients with **iliofemoral DVT** results is less **post-thrombotic symptoms** and an improved health-related quality of life.

Commonly asked questions

What is thrombolysis?

Thrombolysis is a form of treating **blood clots**, which uses a drug called a **plasminogen activator**. The drug activates the body's **plasminogen** to form **plasmin**, which is the enzyme that actively dissolves **blood clot**. **Thrombolysis** is best performed by delivering the **plasminogen activator** directly into the **blood clot**.

Which patients derive the most benefit from catheter-directed thrombolysis?

Patients who have extensive **deep venous thrombosis** benefit the most from **catheter-directed thrombolysis**. These patients usually have **iliofemoral** (pelvic and thigh) **venous thrombosis**.

What are the objectives of thrombolysis or surgical thrombectomy?

Thrombolysis is aimed at (a) preventing **pulmonary embolism** (clot moving to the lung), (b) reducing or eliminating the acute symptoms of extensive **venous thrombosis** (pain, swelling, loss of limb) and (c) reducing or avoiding **post-thrombotic symptoms** (pain, swelling, skin damage and even breakdown of the skin (ulcers).

Are there other benefits of catheter-directed thrombolysis or surgical thrombectomy?

Yes. Other benefits of **catheter-directed thrombolysis** include eliminating **obstruction** (blockage) of the **deep venous system**, potentially preserving the function of the **vein valves** (prevent **valve** damage), and identifying an underlying **stenosis** (narrowing) of an **iliac vein**, which can be corrected with **balloon angioplasty** and/or **stenting**. By correcting the **iliac stenosis**, recurrent clotting may be avoided.

What are the risks of using thrombolysis to remove free blood clot?

The main complication is **bleeding**. The most commonly occurs from the site where the skin in stuck by a needle to place a catheter or from other needle punctures that the patient may have had in other places. Serious intracranial (brain bleed with stoke) **bleeding** is rare in low-risk patients. **Pulmonary embolism** (**blood clots** moving to the lungs) is also a rare complication.

Are there alternatives to catheter-directed thrombolysis for extensive venous thrombosis?

Patients who cannot receive **thrombolytic agents** can be offered a surgical procedure called **venous thrombectomy**, which is an operation designed to physically pull the **blood clot** from the **vein**. High-risk patients who are not candidates for either **thrombolysis** or **thrombectomy** should be offered conventional **anticoagulation** (**blood thinners**).

Why are blood thinner used after thrombolysis or surgical thrombectomy?

Blood thinner are used to prevent the **vein** from clotting again. If the **vein** clots after treatment, the benefits of the procedure are lost. **Blood thinners** are usually used for 6 months. However, if this is the second or more event, or if you clot more than most people for some unknown reason then you may need **blood thinners** for life.

CHAPTER 9
INDICATIONS FOR INFERIOR VENA CAVA INTERRUPTION

Original authors: Lazar J. Greenfield, Venkataramu N. Krishnamurthy, Mary C. Proctor, and John E. Rectenwald

Abstracted by Gary W. Lemmon

Introduction

The majority of **blood clots** that develop in the **veins** (**venous thrombosis**) occur in the **deep veins** of the pelvis and the legs. The standard treatment for **venous thrombosis** is **anticoagulation** using **heparin**, a **blood thinner** given in your **veins** or into the fat of your abdomen, to immediately prevent more clotting and changed to a drug given by mouth (**Warfarin** or **Coumadin**) for the long term treatment. This therapy successfully reduces the potential for life threatening **pulmonary embolism** (**PE**), **blood clots** moving to the lungs, by a factor of over 100 times. Unfortunately not all people are able to follow this standard pathway. Those unable to use standard medicines typically fall into one of four groups. These are:

1) Contraindications (reasons not) to use **anti-coagulant therapy** (**blood thinners**) (example: **bleeding** stomach ulcer).
2) **Bleeding** that starts while using **blood thinners** to the point that **blood thinners** can not be used.
3) New **pulmonary embolism** or more **venous thrombosis** which happens while on **blood thinners**.
4) Individuals having a massive **PE**, **clot** in the lung, that brings the person near death if something more is not done.

Failure of medical treatment plans for the reasons just noted makes your doctor consider other methods protecting you from **pulmonary embolus**. This treatment involves a **filter** (screen) inserted into the biggest vein in your abdomen to prevent **blood clots** moving from the legs into your lungs.

Surgical Treatment Advances

One way to prevent **blood clot** from moving from the legs to the lungs is by tying off the main vein in the abdomen (**vena cava**) but that resulted in too many problems for very sick people. Then methods were made to partly interrupt, by sutures or staples, the **vena cava**. These had better success but still needed an opening of the abdomen in the ill patient to place the sutures or staples. Directly opening the abdomen to work on the **vena cava** was little used after devices called **vena cava filters** could be placed into a distal **vein** and passed into the **vena cava** to do the same thing. With information gained from animal and then human studies, the **Greenfield filter** (a cone shaped device used to catch **blood clots** traveling in the **vein** blood) was the first to offer this protection of decreased risk of **PE** in addition to providing a low risk of death and surgical complications for the patient. Continued improvements in **filter** design have decreased the size of the devices and the easy of placing the filters by x-ray or with ultrasound (sound

Provided by the American Venous Forum: veinforum.org

wave) guidance today. The device used to place the **filter** is typically smaller than a soda straw. When placed in the **vena cava**, the **filter** acts as a screen that allows continued passage of normal blood flow back towards the heart but is able to trap **clots** less than an eighth of an inch in size.

The **filter** placement is done with the patient on an x-ray table. The groin or neck is clean to make it sterile and a needle is used to gain entrance into the **femoral** or **jugular vein**. A small wire is then placed into the **vein** over which is placed a catheter (tube) which can be pushed into the **vena cava** under x-ray or sound wave viewing to the proper place in the **vena cava**. The **filter** is place through the catheter to the correct place and its spring loaded design allows it to rapidly open and stick to the **vena cava**. Once properly placed, the devices used to get the **filter** to the correct place are removed and gentle finger pressure is held over the site of **vein** stick to stop any **bleeding**. The patient may resume normal activity by the next day.

As it became easier to place these **filters**, the number of reasons for **inferior vena cava filter** use has increased beyond the traditional means as mentioned above. Several **filter** designs are now available for clinical use **(Figure 1)**. Most are made of stainless steel or a nickel-titanium alloy. All **filter** designs allow for capturing of **blood clots** from the legs or pelvis. This provides a markedly reduced risk of a catastrophic loss of life from a large **blood clot** causing **PE** and the inability to supply oxygen to the blood. Recent advances have included **filter** placement in the intensive care unit setting using **ultrasound (sound wave) guidance**. Further advancement in design has included the ability to place and then remove the **filters** at a later time after the risk of **PE** has been reduced or eliminated.

While an **inferior vena cava filter** can significantly reduce the risk of a life ending **PE**, recurrent **PE** risk remains in two to three percent of patients. Therefore when possible, continuation of **anticoagulant** medication after **filter** placement remains an important part of the continued treatment for **venous thrombosis** and prevention of **PE**.

Conclusion

Blood clots from the large **veins** of the legs and pelvis can produce life threatening **PE** if left untreated such as when standard therapy cannot be used. **PE** management has evolved over decades and the current state of the art treatment includes placement of an **inferior vena cava filter** which has very minimal risk. While extremely effective in preventing **PE**, it does not reduce the risk to zero nor does it treat the underlying venous clotting problem. Therefore continued **anticoagulant** therapy (**blood thinners**) if possible, should be used when possible.

Commonly asked questions

Is it safe to leave the filter in and does it has to stay there for my lifetime?

Once placed, **inferior vena cava filters** are very safe and well tolerated. Over 92% of vena cava **filters** are present for a lifetime without major problems. The stainless steel and other metals used to make **filters** cause little reaction in the body. They are safe to go through airports and will not trigger metal detectors. Most are safe for x-ray studies and metal alloy **filters** can be

used in Magnetic Resonance scanning devices. Recent filter design advances allow for removal at a later time when the threat of **PE** has been eliminated.

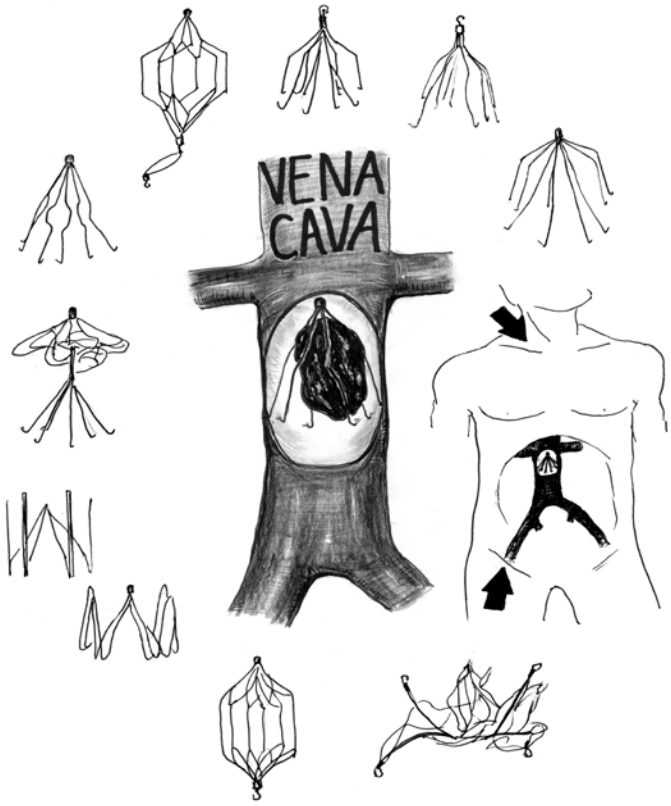
Can the filter be placed for blood clots in the arms rather than the legs?

Yes, the filter can be positioned in an up-side-down position in the **superior vena cava** (biggest **vein** in the chest) to allow for successful trapping of **blood clots** from the arms or the neck. While this is not a common problem, it is not normally needed but when needed can be placed much as one placed a **filter** in the abdomen **vena cava**.

With so many filter types which one is the best for me?

Filter selection is based on what your doctor thinks is best, on your particular body shape and the particular design of each filter. The choice of which **filter** to use is best selected when all these factors are taken into consideration.

Figure 1: Various filters are designed to work much like the spokes of an umbrella without the material on it. They will allow for trapping of a blood clot below the cone of the filter. They work because of the particular flow within the vena cava which funnels the majority of blood clots to the center of the vessel. The advantage of this open metal design allows for continued blood flow through the filter while being very effective at trapping clot.



CHAPTER 10
AXILLARY/SUBCLAVIAN VEIN THROMBOSIS (CLOT) AND IT'S TREATMENT

Original authors: Richard M. Green and Robert Rosen

Abstracted by Gary W. Lemmon

Introduction

Venous thrombosis or blood clots occurring in the large veins draining the arm is termed **axillo-subclavian vein thrombosis**. Some people are prone to developing **venous thrombosis** from muscular and skeletal abnormalities surrounding the **subclavian vein**. Such abnormalities are generally located in the area of the body called the **thoracic outlet** where nerve, artery, and **vein** go from the neck and chest to the arm. Abnormal muscle bands or abnormal rib development can "pinch" the **subclavian vein** creating a severe narrowing as the arm rotates around the chest in its normal range of motion. Those having this altered anatomy may then develop **venous thrombosis** of the **subclavian vein** from repetitive trauma to the **vein** as the arm moves. Unusual and forceful arm motion as happens in pitching or other prolonged labors such as house painting or window washing may cause **vein thrombosis**. When **thrombosis** occurs in the **axillo-subclavian vein**, marked arm swelling occurs with a typical bluish discoloration of the hand and fingers.

Diagnosis

An accurate history and physical examination in addition to the rapid presence of significant arm swelling and tenderness over the upper chest makes the diagnosis, especially when such findings follow the type of activity known to cause **thrombosis**. Appropriate confirming studies can show the **thrombosis** (blood clot) in the **axillo-subclavian vein**. In the past, contrast **venogram** (placing a needle in the **vein** and injecting x-ray sensitive dye into the **vein**) was the preferred study. Presently, the confirming study is high quality **computerized tomography (CT) scanning** or **magnetic resonance imaging (MRI)** studies.

Management

The treatment of **axillo-subclavian vein thrombosis** has undergone change over the years. If no treatment is provided except for arm elevation and pain relief, the patient can have long lasting arm swelling and discomfort due to the venous blockage. The arm remains heavy, stiff, swollen, and generally uncomfortable. **Blood thinners** such as **heparin** or **Coumadin** are part of the treatment for this disorder but these drugs do not actually remove the clot or the abnormal anatomy which is "pinching" the **vein**. Use of a **blood thinner** alone results in chronic disability and persistent swelling and pain in up to 75% of patients.

Better results have been obtained when drugs that are made to breakdown or dissolve the **venous clot** (**thrombolytic agents**) are placed into the **clot**. They are best placed into the

clot by a catheter (small plastic tube) which is pushed right into the **clot**. This catheter is placed into a **vein** further out in the arm and pushed to the place of clotting. It is not uncommon for an overnight delivery of these drugs to achieve complete **clot** removal. While **thrombolytic agents** can dissolve the **clot**, it is recommended to continue **anticoagulation** with **heparin** after clot removal since recurrence of **thrombosis** is common if this is not done. Once the **venous clot** has dissolved, marked improvement in arm swelling occurs. The impingement on the **subclavian vein** must also be taken care of at the level of the ribcage. Treatment with surgery designed to eliminate the "pinch" on the **subclavian vein** is necessary for best results. This typically means removal of abnormal muscle bands and possibly the first rib to free up the entire **vein**. If the vein is scarred from repeated "pinching", pushing the vein open from the inside can be done by placing a dilation balloon and metal support (a **stent**) within the **subclavian vein** during or after surgery. An alternative to **stent** placement can be to directly patch the **vein** open at the time of surgery.

Complete treatment may include **catheter directed drug infusion** to dissolve the **clot**, **anticoagulation** (**blood thinning**) therapy and the surgical removal of the abnormal rib and muscle anatomy. The best results are achieved when performed at the time of diagnosis or by a staged procedure within a short time (six weeks to three months) after clotting. Delays in treatment after initially finding the **clots** or clot removal that does not include eliminating the mechanical pinching of the axillo-subclavian vein has been associated with a higher number of patients with chronic complaints and arm related symptoms that cause disability.

Conclusion

Axillo-subclavian vein thrombosis occurs as a result of abnormal muscle and/or rib anomalies of the **thoracic outlet** at the base of the neck and ribcage. Professional athletes to housewives can be affected. Treatment is best performed close to the time of diagnosis and involves three steps which includes dissolving the **clot**, maintaining **anticoagulation** (stopping any new clots from forming) with a **blood thinner** and surgical treatment to eliminate the external compression on the **subclavian vein**.

Commonly asked questions

What is thoracic outlet syndrome and who can be affected by it?

Thoracic outlet syndrome **(Figure 1)** involves a group of arm symptoms produced by abnormal muscles, scar bands or bones (rib or collar bone) located in the upper ribcage which pinch the main nerves, artery and **vein** of the arm as they leave the neck and chest to get to the arm. **Axillo-subclavian vein thrombosis** (clotting of the major **veins** of the upper arm) affects approximately five to eight percent of all patients with **thoracic outlet syndrome**. When pinching of the **subclavian vein** is the major compression, **venous thrombosis** (**blood clots**) can occur often after long periods of physical activity or unusual movements of the upper arm. Thus, "weekend warriors", housewives with long period of window washing, house painters, weightlifters, professional athletes such

as volleyball players, and baseball pitchers can all be affected by this disorder. Left untreated, chronic fatigue, arm swelling and discomfort are the norm. The common problem affecting all those people involves **subclavian vein** compression and injury.

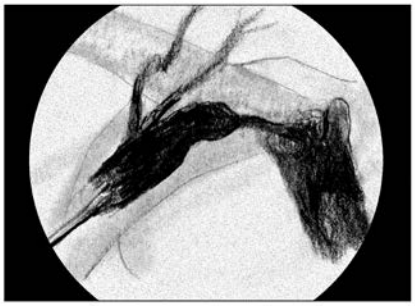
Can both arms be involved?

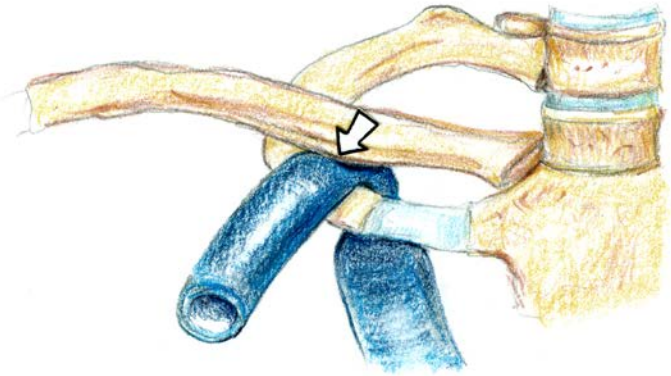
Yes. **Thoracic outlet syndrome** disorders can be on both sides in over 50% of patients. While it is most common in the dominant arm, it can also occur in the non-dominant arm.

What type of disability remains with treatment?

As mentioned, left untreated, chronic arm symptoms such as fatigue, swelling, and aching is common. This can make it difficult for a person to perform their normal activities, sports or job. If promptly treated, recovery is relatively rapid. Normal activity can usually take place in four to six weeks. Chronic **anticoagulation (blood thinner)** treatment with **Coumadin** may be necessary for a long period of time.

Figure 1: The upper panel is an artist impression of a venogram (an X-ray agents injected directly into the major veins of the upper arm and chest) showing pinching of the vein at the thoracic outlet. The lower panel shows the clavicle (upper bone) pinching the subclavian vein as it goes over the first rib to more clearly show what is happening in the upper panel. Successful catheter directed thrombolysis (clot removal) has reopened the vein but the narrowing can easily be seen that requires surgical treatment.





CHAPTER 11
PHYSIOLOGY OF VENOUS INSUFFICIENCY

Original authors: Kevin G. Burnand and Ashar Wadoodi

Abstracted by Teresa L. Carman

Introduction

Chronic venous insufficiency is a broad term which is not well defined. The term is used by doctors for many different venous problems. No matter what term you want to use, chronic venous insufficiency or **chronic venous disease** is seen as **leg swelling**, skin darkening (**hyperpigmentation**), skin thickening (**lipodermatosclerosis**), and when severe enough can cause an **ulcer** (sores or wound) most often seen near the ankle. Normal venous blood movement (circulation) depends on a normal central pump (the heart), a **venous pressure gradient**, a normal **calf muscle pump**, and normal **vein** structures with intact **valves**.

For **chronic venous disease** causing chronic skin changes, the latter two (a normal **calf pump** and intact venous structure) are likely the most important. The leg has 3 muscular pumps (muscle groups that can push blood out of your leg toward the heart): the **foot pump**, the **calf muscle pump**, and the thigh pump. The foot pump "primes" the system or fills the lower leg **veins**, the **calf muscle pump** generates most of the push to drive the blood out of the lower leg and through the **veins** (blood vessels carrying blood out of the leg and toward the heart), and the thigh pump is the least important providing support to the rest of the venous piping. In a normal system, when the **calf muscle** contracts or moves it produces a lot of force which pushes blood through the **veins**, up against gravity and back towards the heart. When the muscle relaxes and the veins lying within open up, it creates a gradient (less pressure within in comparison to the other **veins**) which draws the blood from the **superficial system** into the **deep system** to help with venous blood movement. In a normal person, an intact **calf muscle pump** is efficient – forcing about 70% of the blood out of the calf as we walk. If the **calf muscle pump** and venous piping and **valves** lying within are not working, **chronic venous insufficiency** becomes a problem for the patient.

Chronic Venous Insufficiency

Two other major contributors to **venous insufficiency** are **venous reflux** and **venous obstruction**. There are three different regions (locations) of **veins** (the piping or tubes within lies the blood) within the leg – the **superficial veins**, the **deep veins** lying within the muscles, and the **perforating veins** which connect the **deep** and **superficial veins**. The **valves** are made of two very thin flaps of tissue which meet in the middle to stop the backward flow of blood within the **veins**. If the **valves** have been injured or are not normal, **venous incompetence** or **reflux** can happen allowing the blood to **reflux** or go back down the veins into the legs **(Figure 1)**. **Venous reflux** may occur in the **superficial veins**, the **deep veins** or the **perforating veins**. If the inside of the **veins** (the lumen) is

scarred or blocked (**venous obstruction**), this will prevent the blood from getting out of the leg (prevent normal **venous outflow**). Any or all of these problems can cause **venous hypertension** or elevated pressures within the **veins** which can push fluid and protein out of the **vein** and into the lower leg tissues which then cause skin and tissue changes associated with **chronic venous insufficiency**.

Deep venous reflux or **obstruction** often is seen after a **blood clot** in the lower leg (**deep vein thrombosis**). **Blood clots** may occur whenever there is an injury or trauma to a **vein**, poor blood flow or slow blood flow in a **vein** (decreased venous blood circulation), or when the blood clotting is too active. Once a **blood clot** is formed the body has agents (proteins) which work to breakdown the **blood clot** (called **thrombolysis**) and clean up and restore the **vein** to normal. In about 50% of cases, this happens over about a 6 month period of time. In many people, the **blood clot** is never fully cleaned up and there will remain scarring and damage of the inside of the **vein**. In some cases, there is just a small amount of damage seen as 'webs' left in the **vein** while in other cases there may be a large amount of damage seen as complete scarring and loss of the entire inside of the **vein** causing blockage of blood movement (**obstruction**). The body works to make up for this blockage by opening up smaller **veins** (collateral **veins**) for the blood to move around the obstruction or by making holes in the **vein** which still allow some blood movement in the damaged **vein**. In common terms, if you compare the major **veins** to the highways and the collaterals channels to the side streets – blood still can flow out of the leg and back to the heart but the side streets are never as good with moving the cars as the highways would be. Therefore the amount of the **obstruction** (blockage) and the efficiency of the collaterals (side streets) determine the impact of the **blood clot** on **venous return**.

Along with scarring of the **veins**, a blood clot may injure the **valves** (two leaflets lying in the **vein** which shut to stop the backward flow of blood) causing **incompetence** and **deep venous reflux** or back flow **(Figure 1)**. This will allow the blood to go backward in the **vein** and down the leg rather than up to the heart. In most cases, a strong **calf muscle pump** can make up for this unusual blood flow. However, if the **calf muscle pump** is not working or if there is damage to the **perforating veins**, this unusual backward flow of blood can cause **venous hypertension** or elevated pressures in the lower leg.

When there is **obstruction** or **reflux** following a **blood clot**, it is called the **post-thrombotic syndrome**. Up to 70% of patients may have signs and symptoms of **post-thrombotic syndrome** 7-10 years after a **blood clot**. The changes of **post-thrombotic syndrome** may range from mild **swelling** to severe **skin changes**. In the worst cases, this can cause open wounds known as venous stasis **ulcers**.

Superficial venous reflux unlike the **deep** disease usually happens because the **valves** or **vein** become weak and floppy as a person ages. This may cause the typical **varicose veins** which are easily seen ropey and large **veins** in the thigh or calf. In some people, **superficial reflux** may have no symptoms, may be noted as only very small **veins** (called "**reticular veins**") or even smaller very thin reddish colored **spider veins** (called "**telangectasias**").

Perforating veins connect the **deep** and **superficial veins**. With their one-way **valves**, they allow blood to flow from the **superficial system** into the **deep veins** and prevent back-flow. When the **perforating veins** are incompetent or leaky, this will also increase the pressure in the **superficial veins** and contribute to the **skin changes** of **chronic venous insufficiency**.

Chronic Vein Insufficiency Effects

Regardless of the cause, **chronic venous insufficiency** and **chronic venous hypertension** may result in **leg swelling**, skin darkening or **hyperpigmentation**, skin thickening or **lipodermatosclerosis** (fat and skin scar development), and even sores or **ulcers**. Elegant studies have shown that there are several abnormal changes that happen in the **veins** which cause the **skin changes** seen. There is increased blood flow in the area of the **skin changes**. However, despite an increase in blood flow there is a decreased delivery of oxygen and life-giving food to the tissues. The leaking of red blood cells into the tissues through small holes in the vein walls helps to cause the skin staining or **hyperpigmentation** seen in the disease. Work is taking place to learn what is causing the wound or **ulcer** to form.

Commonly asked questions

Why do my legs swell?

Leg swelling may happen for many reasons. When **venous disease** is the problem, it may be because of increased pressure in the **veins** from heart or lung disease or because the **veins** are not working properly. The **veins** may not be working well because the **valves** are worn out causing backward movement of blood into the leg (**reflux**). Other causes of **swelling** may include liver disease, kidney disease, medications, problems with water balance, or mechanical loss of the "**calf muscle pump**". Your doctor can do laboratory testing and vascular testing to determine what conditions you may have.

What if my swelling just happened and is only in one leg?

Swelling that happens in only one leg may be more likely a problem with blockage or **obstruction** of the **vein**. If it just happened, you need to seek medical attention to make sure there is no acute **blood clot** forming in your leg **veins** (**deep vein thrombosis** (**DVT**)) or other important cause.

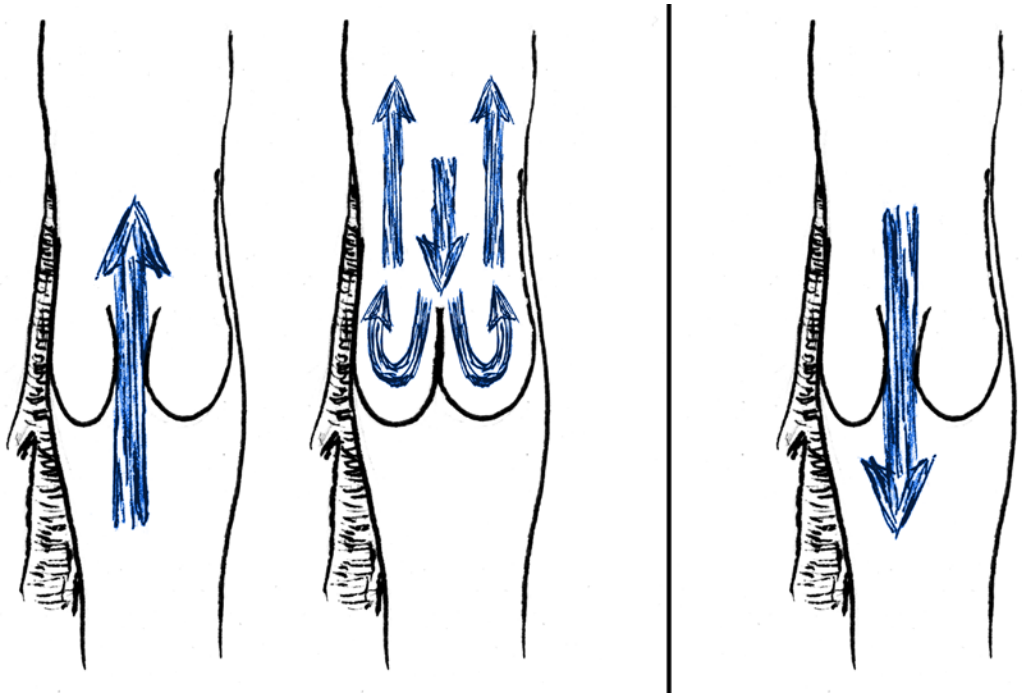
What causes the veins to not work well?

In many cases, **veins** may wear out because of a family trait (inherited) passed on to you. If you have a family history of **varicose veins**, this may not be avoidable. Trauma or injury to the **vein** is another cause for the **veins** to not work well. This is the case after a **blood clot** or **deep vein thrombosis**. After a **blood clot**, many people will suffer from changes related to the **post-thrombotic syndrome**. Your doctor can help to diagnose any of these problems.

What can I do about my swelling and what can happen if I do nothing?

Once your doctor finds the reason for the **swelling** then treatment can be started. In many cases, elevating the foot of the bed to help get the blood out of your leg while sleeping or by using **compression stockings** while awake and upright may be very helpful. Of course, if there is a problem with your heart or other problems your physician will need to take care of these problems also. Keeping the skin moisturized may help with the dry skin changes. You will need to protect the skin of your leg from injury if you start to see thickening of the skin since then it becomes more likely to be damaged with very little bumps or cuts. Open wounds or **ulcer** can be seen usually at the ankle which needs medical attention to heal.

Figure 1: An artist's picture of the workings of a normal valve (first panel with two pictures) where the valve opens with calf movement and then closes to stop the reflux or backward movement of blood when standing still. The second panel show a valve which does not close properly and so allows blood to flow back into the leg when standing and immediately after your stop walking.



CHAPTER 12
CHRONIC VENOUS INSUFFICIENCY PRESENTATION

Original authors: Andrew W. Bradbury, Andrew D. Lambert, Robert B. McLafferty, and C. Vaughan Ruckley

Abstracted by Teresa L. Carman

Introduction

The diagnosis of **venous disease** is difficult to make on taking a simple history of the patient's problem and physical findings (what the doctor sees). For this reason, most patients will have to have more tests done. That being said, many important facts about the patient's problem are seen on physical examination. The doctor's examination of the patient directs what next tests are needed, how the tests should be done and what the findings mean. **Venous disease** may range from simple **varicose veins** which may be quite asymptomatic to severe **chronic venous insufficiency** with associated **ulcers** or wounds.

Incidence

Varicose veins are common; in fact, some people consider them normal since they are so common. The older the people being examined, the more common **varicose veins** are seen. Most people over the age of 60 have some form of **varicose veins**. In many patients, there is a familial tendency (genetic basis) with their mother or father having **varicose veins**. The problem is more common in women than in men. Being overweight, pregnant and/or having to stand for long periods of time may increase the risk of having **varicose veins**.

There are three kinds of **varicose vein**: **varicose veins** of the **saphenous** or just off the largest **superficial veins** (**trunk varices**), **reticular veins**, and **telangectasias** (**spider veins**). The typical **varicose veins** that most people think of are the **trunk varices**. These are directly part of the large **superficial veins** of the leg including the **great saphenous vein** (the largest vein going from groin to ankle in the fat just under the skin) or the **small saphenous vein** (the largest **superficial vein** going from the ankle to just behind the knee). These veins are almost ½ inch in size and may be larger. They can usually be easily felt especially when the person is standing. **Reticular veins** are small blue veins seen under the skin. They are usually < 4 millimeters in size (less than a tenth of an inch). They usually can not be felt through the normal skin. Up to 80% of adults may have **reticular veins**. **Telangectasias** or **spider veins** are very near to the surface of the skin. These **veins** are usually bright red or purple. They are < 1mm in size, so very small. More than 90% of adults have **reticular veins**.

Presentation

Most people with **varicose veins** do not have any symptoms or so little problems that they chose not to seek treatment. Patients who want to get rid of the **varicose veins** are usually unhappy with the look of their legs, they have problems they think are do to the **varicose veins**, or they are worried that they will develop a worse problem if they do not care for the **varicose veins**.

In general complaints noted with **varicose veins** may be seen in 50% of adults. **Symptoms** commonly seen with **varicose veins** include:

- Dull aching or pain
- Heaviness or the feeling of leg pressure
- Swelling
- Tiredness or fatigue
- Restless legs at night
- Nighttime cramping
- Itching or burning

However, these complaints are not only seen in patients with **varicose veins** and are not described as more or less common or worse the larger the **veins** seen on examination (these symptoms are noted related to severity) Also, the worse the **reflux** (backward flow of blood in the **vein**) does not mean that the patient will have worse complaints. (that is, no relationship between the severity of **reflux** on **ultrasound** study and the presence of symptoms). Therefore it can be difficult to know which patients will have relief from their symptoms after surgery or other treatments for the varicose veins. In addition only a small number of patients will go on to have those complaints seen with **chronic venous insufficiency** when they only have **varicose veins**.

Patients with **chronic venous insufficiency (CVI)** develop **skin changes** resulting from high pressures in the **veins** that then affect the fat and skin most often around the ankle. It is seen as chronic swelling, more severe **skin changes** of thickening or **fibrosis** (**lipodermatosclerosis**) or dark color changes called **hyperpigmentation**, or with the most severe condition, venous stasis **ulcers** (open wounds). Usually patients with **CVI** have more than just **varicose veins**. They may have other reasons for the **skin changes** such as high pressures in the **veins** from heart failure or damage of the vessels that remove protein from the leg called **lymphedema**. Severe pain associated with **CVI** is unusual and should make your doctor look for other causes such as poor blood supply to the leg (arterial disease) or infection.

The signs of **CVI** may include: **corona phlebectatica**, **lipodermatosclerosis** or open **ulcers**. **Corona phlebectatica** is a fan-shaped flare of **reticular veins** and **telangiectasias** around the inside of the foot and ankle. **Lipodermatosclerosis** is thickening and **fibrosis** (scar formation) of the skin of the lower leg. This may begin suddenly and be mistaken for an infection or for a **blood clot**. After having **CVI** for a long time, the skin of the lower leg becomes shiny, hard and has a darker color than the surrounding skin. The skin is fixed or anchored to the underlying tissues making the skin tenser and less flexible. They skin may be very dry (**dermatitis**). White scar tissue (**atrophie blanche**) may also be present.

Diagnosis

The search for **venous disease** as an explanation of the patient's complaints should always begin with a good history and physical examination. When obtaining a history your doctor will likely ask you to tell him, in your own words, what problems you are having in your legs; how bothersome the symptoms are and how long they have been present. The doctor will want to know of any other medical problems which might show how important or how likely your symptoms are due to **venous disease**. This includes:

- A history of **blood clots** or other **vein** problems
- Family history of **blood clots** or **vein** problems
- Previous **vein** surgery
- Your job and the need for standing for a long time
- Any issues you have with weight control and constipation
- A history of cancer, stroke, recent surgery illness
- Orthopedic surgery or any injury to the leg.
- When your symptoms happen and if they are getting worse
- The use of compression stockings
- Pregnancies or pregnancy complications
- Any condition which affects the movement of the foot, ankle or leg

In addition, the physician should ask about pain with walking and perform a complete examination of the arterial (vessels which bring blood into your leg from your heart) pulses. Patients with an **ulcer** or wound may be asked about the location of the **ulcer**, the size, what it looks like, whether there are signs of infection, and what treatments have been used in the past.

Some common blood testing may be needed to check on how well your kidneys, liver or thyroid is working and if you have any unusual factors in your blood that can increase your risk of forming blood clots. To help check on the **vein** problem directly, several special clinical examinations may be performed. Most doctors examine patients both laying down and standing in a warm room with good lighting. Standing helps increase pressure in the **veins** and makes them more easily seen. The physician will take note of the location and size of **varicose veins**, **telangiectasias**, and **reticular veins**. Any **skin changes** and/or **ulcers** are noted. Listening with a stethoscope over a **vein** may reveal a noise heard with abnormal blood movement (bruit) which suggests turbulence or increased flow in the area. In an area of previous injury, this may suggest an abnormal connection between the artery and the **vein** also called an **arteriovenous fistula**. If there is a large grape-like cluster of vessels, this may suggest an **arteriovenous malformation** or **venous malformation** (birth related blood vessel abnormality).

Varicose veins are large, ropey and bluish in color. This appearance is due to backflow of blood or **reflux** which causes an increase in pressure within the **veins** such that they get bigger. Palpating or pressing on the **veins** while tapping above or below the **varicose vein** may help to know in which direction the blood is flowing. While not commonly checked now, the **Trendelenburg test** can be used in the office to help locate the site of

reflux. This may be useful if **ultrasound** is not available. The test is performed in 2 parts. In the first part the patient lies flat. The leg is elevated to empty to venous blood then a tourniquet or pressure is applied to stop blood flow in the superficial veins. Then the patient stands and the **superficial veins** are watched to see if they fill with blood. For the second part, the tourniquet is removed or the pressure released and the **veins** are watched to see if they fill with blood. This allows the physician to know which **veins** may be incompetent or not working properly. A **hand-held Doppler** or **ultrasound** machine may also be used to show the same thing. This may be helpful for obese patients, patients with recurrent **varicose veins** after surgery or when the site of **reflux** is difficult to determine.

Additional non-invasive testing may be performed in the **vascular laboratory**. Some **vascular laboratories** perform **plethysmography** testing (volume change measurements of the leg) to evaluate for **obstruction**, **reflux** or **calf muscle pump dysfunction**. Depending on the laboratory the type of **plethysmography** may differ. **Impedance plethysmography**, **strain-gauge plethysmography** and **air plethysmography** are available. Your doctor will be able to tell you which type of test may be the best for you.

Doppler ultrasound is another non-invasive test used. This allows the physician to directly see the **vein** and hear blood flow in the **vein**. This test uses sound waves which can go in to the body, hit the **vein** and be bounced back to detecting part of the machine. The test usually begins with testing to look for **deep vein thrombosis** (**DVT**) or chronic injury resulting from a previous **blood clotting**. Then the **veins** are studied with the patient standing to look for **reflux** (downward flow of the blood) in all of the **deep** and **superficial veins**. Testing may take a long time since for each part of the vein a blood pressure cuff is put around the lower leg and filled with air (inflated) to stop **venous flow**; this is kept in place for approximately 3 seconds. Then while examining the **vein** with the **Doppler**, the cuff is rapidly let down (deflated). In a normal **vein** the **valves** will close quickly and there will be no **reflux**. **Valves** that take more than ½ second to close are not working properly. Most **valves** with incompetence take 3-4 seconds for the **reflux** to stop. In some labs, hand compression of the leg is used to make blood go toward the heart and out of the calf. When this flow stops the time for the **valve** to stop backward blood flow is noted. Greater than one second is abnormal. **Doppler ultrasound** can also help identify abnormal **perforating veins** between the **deep** and **superficial venous systems**. Flow should occur from the **superficial system** to the **deep veins**. If the flow in the **perforating veins** is reversed, (from deep to superficial) this is abnormal.

In some cases, you may need a **CAT scan (computed axial tomography)** or **MRI (magnetic resonance imaging)** to evaluate the **veins** and make sure there is nothing blocking the blood from getting out of the **vein** (**outflow obstruction**). Both of these tests may need the use of contrast or x-ray dye to see the **veins** best so may not be useful in the patient with kidney disease. If x-ray dye is needed than a **vein** must be stuck to allow the dye to be given. Metal devices such as a pacemaker may not be permitted with **MRI** imaging and your physician can determine which test may be required for your clinical condition.

Sometimes invasive tests may be required to best diagnose **venous disease**. This is especially true prior to considering a surgery or procedure to correct a **vein** problem, imaging with a **venogram** may be necessary. A **venogram** uses contrast to directly see the veins with an x-ray machine. To do this test, a vein in the foot, behind the knee or in the groin must be stuck with a needle and a small plastic tube (catheter) placed inside. Where the catheter is placed is determined by which **veins** the doctor wishes to study. Using different techniques and positions, your doctor will be able to see the **veins** and determine the best approach to fixing them. In some cases, **intravascular ultrasound** may be performed along with a **venogram**. This technique uses **ultrasound** technology but the **Doppler** or Probe is on a long catheter placed inside of the **vein**. This allows your doctor to take pictures of the **vein** and any problems from inside the **vein** prior to planning a surgery or other procedure for the **veins**.

Conclusions

In general, any evaluation of **venous disease** whether asymptomatic or causing severe symptoms begins with a good history and a physical examination including laboratory studies. Not all patients will require extensive diagnostic testing. The use of non-invasive or invasive diagnostic testing will depend on the problem being studied and the treatment options that are available.

Commonly asked questions

Do I need to bring anything to my first appointment with the vein specialist?

In general, if you have laboratory studies, prior tests or films these may be helpful to your doctor to see so bring the actual study with you to the visit. It will be necessary to have a good history of both your symptoms and any prior treatment. Any records which you can send in advance of the appointment or bring along with you may be helpful.

Do I need to fast or not eat before my appointment?

Fasting is not required for most **venous imaging**. If you doctor plans on doing a **CAT scan** or an **ultrasound** of your main **veins** in the pelvis fasting may be needed. The doctor will tell you to do so before that visit. If you are having an invasive procedure such as a **venogram** then fasting will be required.

Will I need a surgery for my veins?

Most **venous disease** is not a major problem for the patient and does not require a surgery or procedure. Cosmetic procedures to improve the appearance of the legs may be performed in asymptomatic patients but these are not required procedures. Patients with bothersome symptoms or recurrent **ulcers** may benefit from a procedure or surgery.

CHAPTER 13
COMPRESSION THERAPY FOR VENOUS DISORDERS AND VENOUS ULCERATION

Original authors: Gregory L. Moneta and Hugo Partsch
Abstracted by Teresa L. Carman

Introduction:

The first treatment used for **chronic venous insufficiency** and **venous ulceration** is **compression**. The goals for **compression** therapy are: to encourage rapid **ulcer** healing and prevent a **recurrence**. In most cases, complete healing can be accomplished in under 3 months. Barriers to healing may include: older age, obesity, **venous reflux** or back flow in the **veins**, and underlying arterial disease (poor blood flow into the leg). In addition, long-standing or very large wounds, and patients with a history of previous **ulceration** take longer to heal. The ideal **compression** allows the patient to continue their normal activities – they are able to walk and remain active in their daily lives.

All **wound care** should begin with basic wound management. Making sure the wound is clean and free of infection, good nutrition to support healing, has good blood flow and is not subject to any injury or trauma which is critical for **wound healing**. Your doctor can pick the best wound management to make sure these needs are met. **Compression** therapy is applied in addition to the required dressing or bandage appropriate for the wound. Typical **compression** devices may include: **elastic compression stockings**, **paste gauze boots** (**Unna's boot**) and **multi-layered wraps**. Some patients even benefit from the use of **pneumatic compression pumps**.

Physiology

Based on experimental information, the ideal pressure for leg **compression** is likely 35-40 mmHg. Studies have demonstrated that in patients with severe **chronic venous insufficiency**, **inelastic compression** decreases **venous reflux** and decreases the **venous pressure** when walking. However, the exact mechanism through which **compression** improves the rate of **wound healing** is unknown.

Improvement in tissue blood flow may play a role. There is thought to be a beneficial effect on tissue pressure and swelling. The **compression** should promote a decrease in swelling by encouraging fluid to move into the lymphatic channels (third type of blood vessel that removes other debris than only blood from your soft tissues). In addition, the increased external pressure from the compression should stop further leaking of fluid into the skin and fat of the leg. As the swelling gets better, the lack of extra fluid should improve the skins' ability to receive needed oxygen and food required for **wound healing**.

Diagnosis

Many patients find it difficult to use **compression** therapy. This makes patient education critical to improve **compliance** (patient use) of the compression. Patients should be educated about the underlying changes in the **veins** (**reflux** with backward flow of **venous blood** down the leg)

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which may contribute to **ulcers** and poor healing. In addition, the need for **compliance** with therapy for both healing and to prevent **recurrent ulcers** should be stressed. Approximately 70% of leg **ulcers** or wounds around the ankle may be related to **venous disease**. Imaging of the **veins** with **ultrasound** (sound waves used to see the veins much as one sees a baby in the womb) or **venography** (injection of X-ray agents to help see the **veins**) will make sure that **venous disease** is the cause of the **ulcer**. The doctor should take a careful history to make sure there are no other reasons that might be causing the **ulcer** or making it harder to heal. Arterial insufficiency also called **peripheral arterial disease (PAD)** is decreased blood flow into the legs which would mean a poor delivery of oxygen and nutrients to your leg and must be excluded as a cause. When arterial disease and **venous disease** present together which can happen, there may be an even more noted delay in healing and the help given by **compression** may have to be changed for best results. If the arterial disease is severe enough, **compression** may be dangerous even cause gangrene or loss of the leg. Other medical problems that may affect **wound healing** include: diabetes, poor nutrition, or a decreased ability to fight infections. In some patients, **compression** may cause a lot of fluid to be pushed back to the heart and result in heart failure. Patients with these medical conditions should be followed closely.

Forms of compression

While **compression** garments are well studied in **wound healing**; they have also been shown to improve patient quality of life. Symptoms of pain, swelling, skin discoloration, cosmetic appearance of the leg, increased activity levels, improved sleep and decrease in depression symptoms have been documented with the use of **compression** therapy when used in the right cases. No studies have demonstrated one form of **compression** to be better than another with respect to **wound healing**. Successful compression is determined by the ability of the patient to use the therapy as directed by the doctor.

Gradient **elastic compression stockings** (more compression in the foot and less as one goes up the leg so that **venous blood** is pushed toward the heart and not pushed into the foot and lower leg) are the typical medical support hose that most people have come to know **(Figure 1a)**. They are available from several companies in a variety of fabrics and materials, strengths, lengths, and designs. Some stockings are fitted with a zipper to help in stocking placement **(Figure 1b)**. They can also be ordered “custom-made” for people with difficult to fit legs. **Compression stockings** are usually less bulky and likely more comfortable for most patients compared to other forms of **compression**. In addition, with the use of **compression stockings** the patient may continue to wear their own footwear. These types of stockings are often used before experiencing a venous **ulcer** to maintain health skin and combat the effects of abnormal **venous pressure** from **venous reflux** or **obstruction**. They are also the most common type of compressive device to prevent a **recurrent venous ulcer** after healing as occurred.

While the benefits related to **compression stocking** for **wound healing** are well known, there are a number of reasons or actions that will cause them to not do the job. If the patient can not put the stocking on, will not wear the stocking as shown to do so, or can not afford the stocking (insurance will not cover that cost); the results may not be as expected. **Compression stockings**

are “operator dependent” – meaning they only work when they are worn. Poor **compliance** (not used as directed) is a major reason many patients fail to heal their wounds.
Patients with significant pain related to the **ulcer** may not be able to use the higher **compression** levels because of pain. Such patients may be started with a lower grade (less pressure applied) of **compression** and increased as the pain they experience becomes less. Elderly patients who have difficulty putting the stocking on or patients known to be non-compliant do not do as well with this therapy. Obese patients and patients who have difficulty moving or bending following a joint replacement or with arthritis may be dependent on a family member or a other helper to assist with applying the stocking.

There are a number of devices available to help get the stocking on. Open toe stockings may be applied using a silk sleeve **(Figure 2a)**. The sleeve is removed following placement of the stocking. Some patients may benefit from the use of a wire frame – or stocking “Butler” to assist with placement **(Figure 2b)**. Other similar devices are available and may be helpful for some patients.

Unna’s boots or **gauze paste boots** are typically a 3 to 4 layer dressing which is applied by a trained medical professional. The gauze bandage which is loaded with zinc, calamine, or other moist substances is applied from the toes to the knee. Additional layers of gauze may be placed and then an outer **elastic compression** wrap is put over all this material. This must be changed weekly and sometimes 2-3 times a week if the wound has a lot of drainage. Once applied, it requires little care from the patient. Care must be taken not to get the **Unna’s boot** wet. Because it may be bulky, it can be uncomfortable for some patients. Some patients may have to use a larger shoe because of the added thickness of this form of **compression**.

There are other types of three and four-layer compression bandages which may have some added benefit in the care of a particular wound. Some doctors feel that the many layers of **compression** result in a more even **compression** up the leg. The added layers may also provide better absorption of the drainage from the wound. Your physician is responsible for determining what materials should be used for **multi-layered compression** devices. Depending on the materials and the elastic parts, the pressure on the leg may be quite variable from one application to the next.

The stiffer and more **inelastic compression** devices have been shown to have more benefit with respect to pushing the venous blood out of the leg (**venous return**). In some patients, padding is put directly over the wound to provide even more **compression** in that area. Padding may also be required for areas where there is rubbing or friction especially at the ankle crease.

Walking exercises are very important to get the best results from any form of **compression** therapy. Some patients experience a fairly significant decrease in swelling shortly after the device is applied. This is due to the increase in pressure and rapid decrease in **edema**. If this occurs, the **multi-layered bandage** or **Unna’s boot** should be reapplied to have the best pressure to continue seeing a decrease in swelling and better healing.

There is a specialized device consisting of multiple, rigid, Velcro-adjustable compression bands which are designed to go around the leg **(Figure 2c)**. This provides inelastic, rigid adjustable **compression** which can be fit to the leg depending on the degree of swelling. This may serve as a useful alternative for patients who are unable or unwilling to wear a **compression stocking**.

Pneumatic compression devices have been used for the treatment of **lymphedema** and **venous ulceration**. While they are not widely used, these devices may be particularly helpful in people with severe **edema**, morbid obesity, or severely impaired movement in whom other options may be limited. These devices use several cuffs filled with air that can be placed on the leg from foot to thigh and that can be inflated and deflated to pump the venous blood and extra liquid out of the leg. For the best results, these should be used along with good standard **wound care**.

Conclusion

The abnormal backward flow of blood in the leg **veins** or blockages to the blood getting out of the leg can lead to problem including swelling, **skin changes** and even **ulcers**. There are studies which make sure that the **veins** are the problem and no other problem has a major role. Once the diagnosis is made, placing **compression** from the outside of the leg can correct many of the underlying problems such that the symptoms get better. **Compression** therapy comes in many forms and some devices are better for healing **ulcers** (open skin lesions) and other for maintaining a steady state within the leg. **Compression** is the first treatment generally used to correct problems seen with lower leg **venous disease**.

Commonly asked questions

Compression stockings are hard to get on. Do I have to wear them every day?

In order for **compression** therapy to give the best result the doctor wants, it needs be placed on everyday before you get out of bed. A hot shower or having your legs down even for a few hours may promote swelling and make the stockings harder to get on.

How can I make the compression stockings easier to put on?

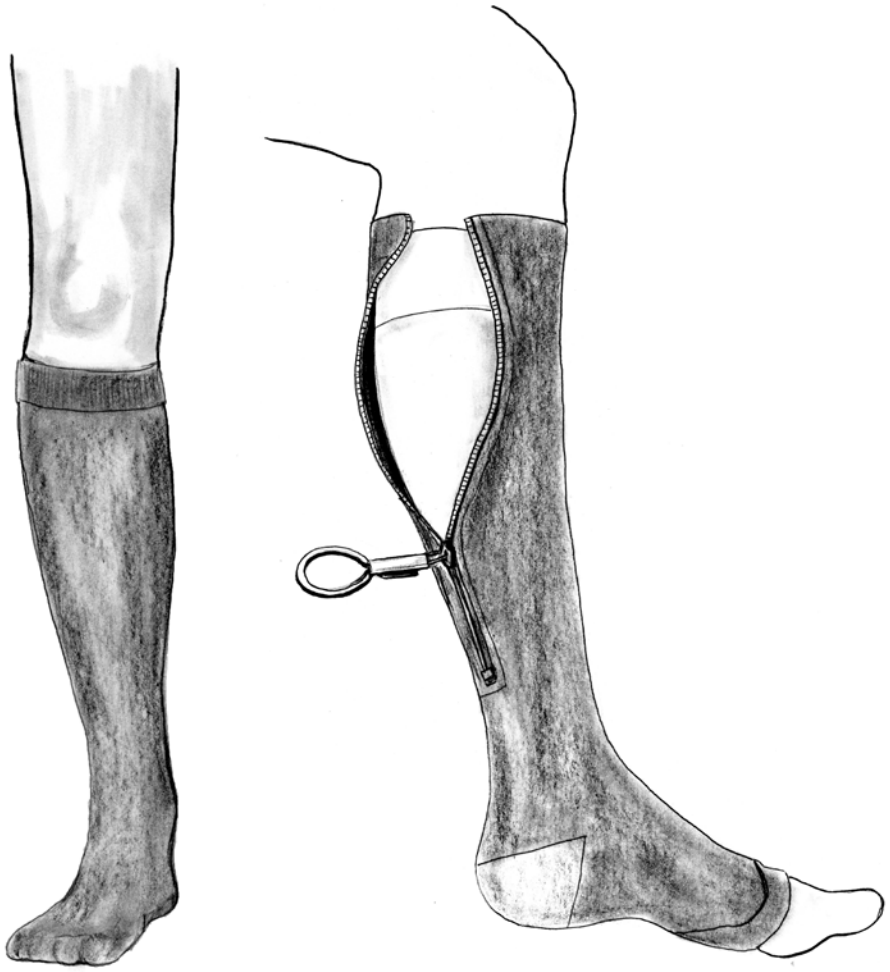
Make sure to put the stockings on first thing in the morning before your legs begin to swell. Moisturize your legs at night rather than just before putting the stockings on since fresh moisturizer may make the skin sticky and more difficult to pull the stockings up. Wearing rubber gloves, like dish washing gloves, usually makes it easier to grip the stocking for the pull up the leg. Use of special aids may be helpful for some patients.

Which stocking is the best?

Stockings come in many different types and styles. For the most part; the material, the color, the style, and the brand of stocking are not as critical as is the proper measurement and fitting. Unless your physician has a particular recommendation regarding a brand or type of stocking – it is most important that the stocking you purchase is the appropriate grade of **compression**

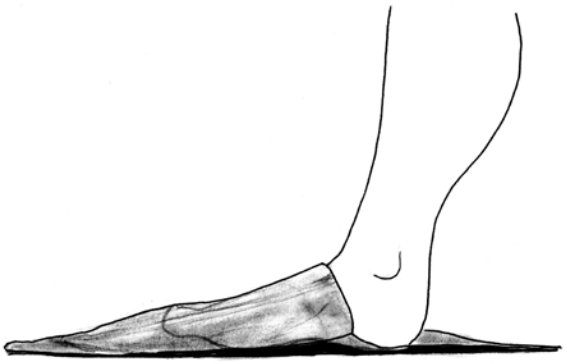
recommended by your physician and that it is measured and fit correctly for you by someone with training to do it properly. Specific characteristics such as a wide band at the top of the stocking or material components which may make donning easier may be recommended by your doctor.

Figure 1: This is an artist's impression of a compression stocking: **(a)** the compression stocking looks much like a normal stocking but is tighter and has more compression in the foot and less at it comes up the leg, **(b)** in some cases, the stocking is fitted with a zipper to make it easier for the patient to get it on.



(a) **(b)**

Figure 2: These pictures are an artist's impression of **(a)** a silk inner which may be fitted on the toes and allows for easier pulling up of the compression stocking and **(b)** the "butler" device which holds the stocking open so the patient can step into the stocking while pulling up the device to "don" it more easily. The Velcro device **(c)** is a series of non-elastic bands which are applied to the leg and adjusted to provide best compression. It can be replaced during the day to have the best compression possible with the least discomfort especially in people who have a tough time putting a stocking on.



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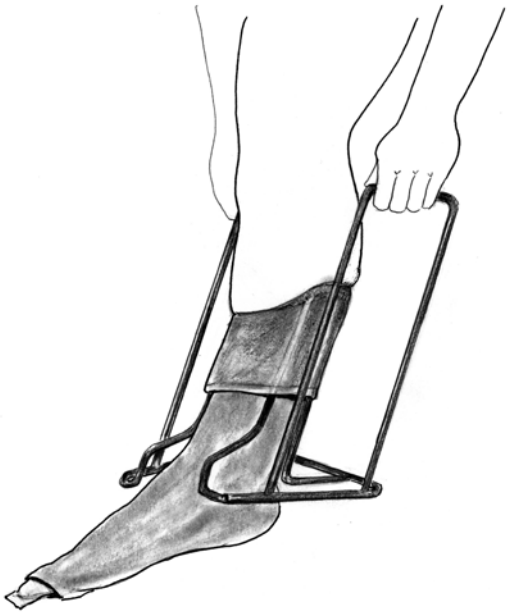

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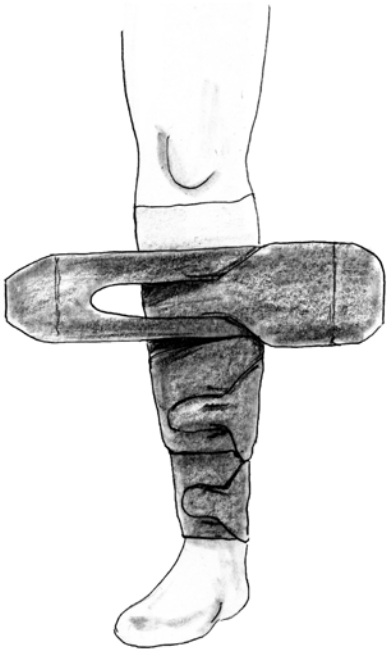

(c)

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CHAPTER 14
SPIDER VEINS/TELANGIECTASIAS THERAPY

Original authors: Jose I. Almeida, Thomas M. Proebstle and Jeffrey K. Raines
Abstracted by Michael C. Dalsing

Introduction

Telangectasias are the medical name for what most people call **spider veins**. These very small **venules** are located just under the skin (in the dermis) and, therefore, are easily seen. There color is often red with or without a predominant bluish tinge. The size is less than one millimeter or 0.039 inches in diameter. The pattern is like a starburst or the limbs of a tree (arborizing).

Over 50% of adults have one or more leg **telangiectasias** and women are most likely to seek treatment to eliminate the blemish (cosmetic reasons).

Etiology and Diagnosis

One cause is an abnormally functioning deeper **vein** with **reflux** or backward flow of blood on standing or sitting. The resulting high pressure in the skin **venules** can cause them to enlarge into a cosmetic blemish. This is important since the failure to recognize and treat the underlying cause can result in the inability to eliminate the **spider vein** for any significant period of time. The diagnosis of lower leg **venous reflux** is made by physicial examination and confirmed by a **venous duplex** study (pictures and blood flow using sound waves) as the first study. There are many other causes which must be considered including congenital and secondary conditions such as exposures (chemicals, radiation), blunt trauma, and autoimmune disorders (lupus, dermatomyositis, etc.). The diagnostic studies required for each are outside the scope of this review, however, your doctor will know best how to determine the cause of the spider veins and the best treatment.

Treatment and Results

If an underlying cause of the **spider veins** is found, its treatment should be completed first to resolve this component of the problem. When the **spider vein** becomes the focus of treatment, generally for cosmetic reasons, the therapeutic use of light energy is a possibility. **L**aser stands for **L**ight **A**mplification by **S**timulated **E**mission of **R**adiation. It uses high powered electromagnetic radiation (focused light) of one wavelength focused to heat one particular small spot of our skin and damage the underlying **spider vein**. Although it would seem so easy to wand away the blemish, in fact, there are many things which have to be accounted for to obtain the desired result. A partial list of important things to consider includes the amount of dark pigment in your skin (melanin), the size of the **venule**, the size of the light beam, the amount of energy given to the skin, scatter of the light when it hits the skin, potential skin damage and pain to the patient. Your doctor

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will consider each before choosing the correct **laser** to use. As an example, the deeper the doctor needs to penetrate the skin to hit the **spider vein** the longer the wavelength needed. Generally, shorter wavelengths (500-600 nm) will only go about 0.7 mm deep (e.g. KTP laser, flashlamp pumped dye laser) while the longer wavelengths (1064 nm, Nd/Yag laser) will go up to 3 mm (one tenth of an inch or more) into the skin. The doctor will cool the skin during laser treatment to prevent burning (to prevent sunburn-like problems). Most doctors will not treat tanned skin because the increase in a certain skin pigment (melanin) increases the absorption of the laser energy leading to tattooing. There are other devices using **intense pulsed light (IPL)** which are different from the **laser** in that the light source is neither one wavelength nor as controlled but otherwise works much like the **laser** when it comes to treatment.

How is it done? In general, short bursts (in the millionth of a second range) are given to lessen the discomfort associated with the treatment. The spot size hit and thereby treated by the light is often much less than one half inch in diameter. Larger areas of the skin are treated by treating different spots without trying to overlap. It usually takes two to four visits to complete the job which, of course, depends on the number of spider veins being treated. Most reports on the **laser** treatment of **spider veins** shows 75% to 100% clearance of the abnormal color and distention of the **spider veins** noted before beginning the treatment. As with every treatment there can be problems (complications). The most frequently reported problems include a worse darkening of the blemish (**hyperpigmentation**), incomplete removal of the blemish, and treatment related pain. Clots with the **spider veins** have been reported as well as burns of various degrees.

Spider veins can also be treated by injection of drugs that will scar the venules and cause them to be less visible. Most doctors use a very small needle inserted into the **spider vein** to inject the damaging drug which might be very concentrated salt water or other **sclerosing drugs (e.g. sodium tetradecylsulfate)**. Pressure is held over the area of injection with the doctor's hand to begin with and then continued with pressure stockings for several days afterward. It works by causing the lining cells of the **venule** to swell and rupture and the surrounding cells to become inflamed and to eventually scar so that the vein is no longer seen. Results are comparable to those noted for laser treatment. Complications are generally local including local **blood clotting** and **ulcers** from the drug leaking out of the vein. However, allergic reactions (blood pressure, heart and breathing problems) are also a rare possibility.

Conclusions

Spider veins are very small blemishes within the skin. The cause of the **spider veins** must be sought and treated prior to taking care of the skin blemish. To eliminate the **spider veins**, currently two treatments are commonly used. **Laser** treatment uses light to heat the **spider vein** resulting in scarring while **sclerotherapy** uses drugs to damage the inside of the **vein** resulting in scarring. Each method has risks including a worsened cosmetic appearance for the potential benefit of eliminating the blemish.

Commonly Asked Questions

What are spider veins?

Spider veins are very small reddish blue venules located in the skin which have enlarged and become visible.

What causes spider veins?

Spider veins can results from minor skin trauma which damages the small skin **venules**, from dermatologic or autoimmune disorders, from exposure to damaging agents, or from underlying **venous reflux** with resulting high blood pressure in the **venules**, enlargement of the **venules** and ultimately the visible blemish one sees.

Are there ways to get rid of my spider veins?

Yes but first one must make sure that the **spider vein** is not a sign of some deeper problem such as **venous insufficiency** (**reflux** in larger **veins**) or other medical problem. The **spider veins** can be made pale and shrunken (less visible) by using **laser** energy or **sclerosing drugs** to damage and scar the **spider vein**. **Laser** energy is delivered by shining the light on the skin while the **sclerosing drug** is injected into the **vein** with a very small needle.

CHAPTER 15
SCLEROTHERAPY FOR VENOUS DISEASE

Original authors: Niren Angle, John J. Bergan, Joshua I. Greenberg, and J. Leonel Villavicencio
Abstracted by Teresa L. Carman

Introduction

New technology has changed the management of **venous disease**. Treatments using a catheter placed inside the **vein** (**endovenous therapy**) to deliver radiowaves (**radiofrequency**) or light energy (**laser**) therapy to the inside walls of the **vein** are now used in addition to traditional surgical **vein stripping** for **varicose veins**. **Sclerotherapy** using liquid or foam agents is one treatment not only for some **varicose veins** but other types of **venous disorders**. **Sclerotherapy** involves the injection of a drug into the **vein** to cause irritation to the inside of the **vein**. This irritation along with **compression** of the **vein** causes the **vein** to scar closed. **Sclerotherapy** may be used alone or along with surgical treatment to remove the **varicose veins**. Most physicians are familiar with all of the available ways for treating **varicose veins** and should able to decide on which is best for your **veins**. In many cases, more than one technique is used.

Treatment of **venous disease** begins with knowledge of the best tests to determine which **veins** would benefit from therapy, the indications for the procedures, and an understanding of which patients will not benefit from therapy. In addition, your physician should be familiar with the complications or problems related to the treatment that may occur and know how to manage these complications. Prior to the treatment, the patient should be told what therapy that is recommended and way, what results to expect both immediately following the procedure as well as what to expect in the weeks to months after the treatment, and any complications that may happen. Photographs are usually taken before the procedure. A consent form is used to make sure you have had the opportunity to ask questions about the procedure.

The first visit with the patient begins with a good history of what has been happening with the patient to help decide on how bad the problem is, what may have caused the problem: for example a history of **blood clots** or **vein** injury, and any other medical problems or medications that may affect the choice of treatment. The physical examination should be performed in a standing position. Most doctors will note and some will take pictures of the venous problems. This includes **varicose veins** but also smaller **reticular veins** and "**spider veins**" or **telangectasias**. Other skin changes or findings may also be important such as patients with skin injury (color changes or skin thickening) or swelling related to the **veins**. Some patients will need blood work before the procedure to make sure they are not at risk for other complications. If the **varicose veins** were caused by a **blood clot**, a hematologist or someone who specializes in **blood clotting** problems may help in the care of the patient. Most physicians will use **ultrasound** to help them determine which veins are not working well and therefore need to be treated. If the

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varicose veins are not typical such as **venous malformations** or are going into the abdomen and not confined to the legs then more extensive testing may be needed.

All patients with **venous disease** will have their symptoms helped by elevation of the leg, external compression using special stockings, and good skin care. If possible, you should elevate the foot of the bed 3-4 inches by placing a wooden block or brick under the bed post. This improves blood getting out of the leg by using gravity to help drain the blood from the legs while sleeping. Clean and moisturize the skin every day to keep it soft and less likely to crack or be injured. These simple steps should be used no matter what other treatment is planned.

Indications (reasons) for using sclerotherapy include**:**

- Treatment of **spider veins** or other skin blemishes
- Treatment of **reticular veins** or small **varicose veins** (1-3mm) when there is no major **reflux**
- Treatment of **veins** < 3 mm that remain after surgery or larger **veins** 3-4 mm that are not due to underlying **perforating vein** problems.
- Treatment of **perforator veins** with incompetence or back-flow in some cases
- Treatment of bleeding **varicose veins**
- Treatment of large **varicose veins** hidden below a **venous ulcers**

Contraindications (reasons not to use) to sclerotherapy:

- Pregnancy (unless associated with bleeding from a varicose vein)
- Age over 75 years in some cases
- Sedentary (very inactive) patients who may be at increased risk for **deep vein thrombosis**
- Patients with underlying medical problems including diabetes, kidney problems, liver disease, cancer, heart disease, lung disease or bleeding problems
- Patients with arthritis that keeps them from walking and enjoying normal activities
- Peripheral arterial disease (PAD) or decreased blood flow to the legs
- Severe allergic reactions or a history of asthma in some cases
- Fever or an acute illness
- Recent or acute **phlebitis (vein infection)**, **superficial vein thrombosis** (**blood clot**) or **deep vein thrombosis**
- **Veins** that are connected to major veins (**saphenous veins** that go up and down the leg and carry most of the venous blood out of the leg) that have **reflux** (incompetence) are likely to recur

Patients taking aspirin or anti-inflammatory drugs like Motrin ® or Advil ® need to stop taking these drugs for 1 week before treatment. Patients on **anticoagulants (blood**

thinners) like **warfarin** are usually not offered **sclerotherapy** until they can be off these drugs.

How is sclerotherapy performed?

Sclerotherapy is performed using different chemicals and are placed directly into your abnormal **vein** through a very small needle. All of these chemicals cause injury to the cells lining the inside of the **vein** and create inflammation and irritation within the **vein**. This allows the **vein** to scar closed. Your doctor will decided on which chemical is best for your condition. In some cases, lidocaine (similar to Novocain®) may be added to the chemicals to decrease the discomfort of the injection. Some physicians will create a foam form (drug mixed with air) of the chemical by shaking it up. This may be especially helpful for larger **veins**.

Sclerotherapy should be performed in a warm, well lit room. The skin is cleaned very well with an antiseptic to prevent infection. A small needle is stuck into the **vein** to be treated and the chemical is pushed into the **vein**. The biggest **veins** are treated before the smaller **veins**. Treatment usually begins in the thigh and moves toward the calf and ankle. When treating **spider veins** usually the most bothersome areas are treated first and the less worrisome areas can be treated later.

Immediately after the injection the **veins** may appear red and swollen or angry. This is due to the inflammation from the injected chemical. Each area is compressed after the injection by taping gauze or cotton over the area. Following the **sclerotherapy** session, **compression** will be applied over the entire leg. This may be done by either a thigh high **compression stocking** or by an elastic wrap. If an elastic wrap is applied you, will need to replace it regularly and should be taught how to do this during your visit. It is important to wear the stockings or elastic wrap as instructed. The **compression** keeps the walls of the **veins** together to allow the scarring to take place and prevent the **veins** from opening again. It also prevents the trapping of blood in the treated **veins** and decreases the risk for pigmentation (unwanted color change). **Compression** is required for a minimum of one week and up to 3 weeks may provide a better result. You should start normal activities and avoid prolonged periods of sitting for the week following treatment.

Complications (problems) of sclerotherapy:

Complications of **sclerotherapy** can occur immediately at the time of treatment or several days to weeks after therapy. There may be local problems confined to the area treated or be more wide-spread (systemic). If the chemical is accidentally injected into an artery instead of a **vein**, this may cause significant tissue injury including **ulceration** or skin breakdown. Your physician should take care to make sure this does not happen. In some cases, **ultrasound** can be used to direct the needle into the **vein** to help avoid this complication.

Local effects which are to be expected include pain, swelling, and redness. This is due to chemical irritation. Allergic response may be experienced which would increase these symptoms and may be associated with itching or development of a rash. In many cases, these symptoms respond to the use of **compression**.

Even when good **compression** is applied after therapy up to 20% of patients will have blood remain in the **veins**. If this happens, the **vein** may develop a brown color as the blood begins to breakdown. In most patients, this brown stain will fade over a period of a few years. Some patients will have small amounts of trapped blood or coagulant in the treated **veins**. This is usually treated with **microthrombectomy** - using a small blade or needle to create tiny holes along the **vein** and remove the blood and lessen the chance for brown staining. This is usually done 2-3 weeks after the **sclerotherapy** injections. After **microthrombectomy**, **compression** is applied to the treated area using gauze pads and a bandage followed by the use of the **compression stocking** or elastic wraps as before.

Bruising (ecchymosis) is common (up to 20%) following **sclerotherapy**. It usually fades over 2-3 weeks after treatment. Older patients and people with frail skin seem to be at increased risk for bruising. **Matting** is the development of very fine red colored blood vessels that appear around a treated area. Between 10 and 30% of patients treated for **spider veins** and up to 15% of patients treated for **reticular veins** may experience **matting**. Obesity and hormone therapy seems to make people more prone to **matting**. Many times this will resolve within a year. Occasionally this can be treated with **laser** light therapy to improve the appearance. Skin **ulceration** or a sore may form at the injection sites. This is usually do to a high concentration of the chemical, injection outside of the **vein**, or injection into a small arteriole. Fortunately these are usually small and respond well to good wound care.

Superficial vein thrombosis or "**phlebitis**" is due to the inflammation created by the injected chemicals. This may occur even a few inches away from the injection site. In many cases, this can be treated conservatively with **microthrombectomy**. In some cases use of warm, moist compresses may be necessary. **Deep vein thrombosis (DVT)** is unusual but has been reported. This is most commonly seen with injection of a large amount of the chemicals or in the patient at risk for prolonged bed rest, with prolonged sitting or if the patient is unable to resume normal activities after the procedure.

In rare cases, patients may experience wide–spread effects such as changes in their vision including bright lights or flashing lights, migraine headaches, dizziness or elevated blood pressure. These usually occur with more extensive procedures which require the injection of large volumes of the chemical agents. In even more unusual circumstances, patients may have a severe allergic response to the chemical agents which can result in difficulty breathing, low blood pressure and may affect the heart beat. Most offices are well prepared to deal with such emergencies.

Foam Sclerotherapy

The technique of creating foam (some type of gas (air or other) mixed with the drug) from the chemicals used for **sclerotherapy** deserves special mention. This technique is gaining popularity and your physician may recommend this as the primary therapy or to help with clearing larger **veins** after a surgical therapy. Because the foam maintains better contact with the **vein walls** it allows the physician to treat larger **veins** more successfully. Typically a smaller amount of the chemical is required which may also decrease the risk for complications. Preparation for **foam sclerotherapy** is the same as for all other forms of therapy. The **vein** is usually entered while looking at it with an **ultrasound**. After the injection the leg is elevated to allow the foam to move into the **veins** down the leg and into any **perforating veins** that are incompetent or refluxing. **Compression** is required after **foam sclerotherapy** similar to all other vein treatments (see above). Contraindications to **foam sclerotherapy** are similar to traditional **sclerotherapy** except that patients with a history of migraine headaches may experience temporary migraine aura symptoms. In addition, patients with a patent foramen ovale (PFO) which is a connection between the right and left chambers of the heart may have a risk of the foam going across that opening and going to the brain. Foam therapy should probably be avoided in these patients.

Complications are similar to that of traditional **sclerotherapy**. **Foam sclerotherapy** may be a bit more painful than traditional **sclerotherapy**. It appears that temporary visual changes are more common with **foam sclerotherapy** than with **liquid sclerotherapy**. This is especially true in patients with a history of migraine headache. One concern arises regarding the entry of foam into the deep vessels at the level of the groin. Since, in most cases, **ultrasound** imaging is being used during the injection the physician is frequently aware of this happening. Foot and ankle exercises will help move the foam from this location and help break up the foam bubbles in an effort to minimize complications. Because of this, the risk for **deep vein thrombosis (DVT)** may be higher with this technique; however, limited clinical studies have not reported this complication to any higher degree. Some of these complications may be decreased by keeping the leg elevated for a minimum of 10 minutes following injection. Despite these recommendations, any visual disturbances, neurologic symptoms, or any unusual symptoms of pain or swelling of the leg after the procedure should be reported to your physician immediately.

Commonly asked questions

Do I need surgery for my veins?

Physicians skilled in the evaluation and treatment of **vein** problems will be able to provide you with all of the information you will need regarding the best treatment options for your condition. Therapy is very individualized to a single patient. The information provided here is to serve as a guideline regarding one possible treatment (**sclerotherapy**) and what to expect. Feel free to discuss details and expected outcomes with your **vein** specialist.

Will I need only one treatment or more?

Most patients will require several **sclerotherapy** treatments to fully treat their **venous disease**. In general, the larger **veins** are treated first and allowed to heal to determine which of the small **vein** need to be treated. You should plan on a minimum of 3-4 treatments to complete the therapy. In addition, many patients will have **veins** return over a period of time and these may require further therapy.

Will my insurance pay for this therapy?

Each insurance company is different but a lot of venous work is considered cosmetic and may not be covered. For symptomatic patients, insurance companies typically require a period of conservative medical therapy prior to authorizing this work to be done and may or may not pay for **sclerotherapy** so you must check with your company.

How long will I be off work after the procedure?

Following **sclerotherapy**, most patients are able to return to work the same day. You need to wear the **compression stocking** or wrap as instructed for a minimum of 3 days and usually as long as 7 days. You should try to walk and avoid activities which prevent movement of the legs such as bed rest or prolonged sitting. You will usually have a follow up with your doctor 7-14 days after the procedure.

CHAPTER 16
SURGICAL THERAPY FOR CHRONIC VENOUS INSUFFICIENCY

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Abstracted by Gary W. Lemmon

Introduction

Varicose veins have been documented from before 2000 B.C. Egyptian papyrus writings trace the use of **compression** bandage to treat **venous ulcers** from the time of the Pharaohs. Treatment for **varicose veins** can be found in writings in 1465 A.D. in a Turkish surgical text. The modern surgical treatment of **varicose veins** started with Frederick Trendelenburg in 1891 when he showed how to tie off the **greater saphenous vein** (largest vein outside the muscle and located in the fat just under the skin) to prevent an abnormal backward flow of blood into the lower leg. The normal method of **vein stripping** (actually pulling the **great saphenous vein** out of the leg) started in the early 20th century by Keller and Mayo and changed very little for many years. Recently, a new way of stopping flow in the **great saphenous vein** to prevent backward flow of blood involves destroying the **vein** while leaving it in place. The procedure is **venous ablative surgery** and uses heat (**laser** or sound waves (**radiofrequency**)) or strong burning drugs (**sclerotherapy**) to destroy the **saphenous vein** without its removal).

Definitions

The normal way the **saphenous vein** works is by allowing blood to flow from the lower leg to the heart (a simple pipe or conduit use) in addition to having **valves** that stop the backward flow of blood once it is push forward by muscles or gravity. This provides a one way direction for blood flow out of the leg. It prevents puddling of blood in the leg which would otherwise be the normal effect of gravity when a person stands. One can well image the swelling that would happen in the leg if such a plumbing system was not in place. Unfortunately, **valves** can become damaged after injury with the result that blood flow backward in the pipes, pools in the lower leg and causing swelling as well as other problems. The term used by doctors for this abnormal state is **chronic venous insufficiency (CVI)**. One other sign that there is abnormal backward blood flow in the lower leg is the presence of **varicose veins** which are often seen as dilated **vein** branches coming off the **saphenous vein** easily and seen beneath the skin **(Figure 1)**. They can be seen as distended irregular bluish colored **veins** going down the leg. Other symptoms seen with **varicose veins** and **CVI** are leg aching, throbbing and progressive swelling in the calf and ankle. **Varicose veins** are often considered unsightly from a cosmetic point of view. If no treatment is provided, the spectrum of symptoms can be very little to pronounced with more severe stages of **CVI** leading to bad skin damage and even **ulcers** (open wounds) usually seen above the ankle on the inside of the leg.

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Economics of Chronic Venous Insufficiency

CVI and **varicose veins** are the most frequent blood vessel abnormalities occurring in people and affects approximately one third of the population. Both men and women are affected and in near equal number (about forty percent of men and thirty percent of women). Twenty to forty year old people in the peak of their productive lives are first burdened by the disability produced by **venous insufficiency**. The result can be lost time from work or from their otherwise active lifestyles. It has been estimated that ten percent of the people are stopped from active employment at some time during their treatment for **CVI**. **Venous disease** has been projected to account for up to two percent of the total health care cost in the United Kingdom alone.

Presentation

The typical patient arrives in the doctor's office with complaints of leg symptoms that include pain, swelling, throbbing and fatigue with or without the presence of a leg **ulcer**. The duration of symptoms is usually progressive from several months to years with the severity of symptoms reflecting the length and degree of **CVI**. A complete history and physical is necessary to identify unusual signs of **varicose veins** and **venous insufficiency**. The history should document any prior **venous thrombosis** (**blood clotting**), history of previous leg **ulcers**, and occurrence of symptoms in other family members and if female, the number of pregnancies and whether **varicosities** near the vagina exist. Additional pertinent history should include current medications, smoking history, and any potential drug allergies. This information is vital if consideration for surgical treatment becomes necessary.

Examination

The patient is best examined in the standing position with the leg slightly flexed and body weight supported by the opposite leg. Feeling for any bulging of the **vein** at the level of the groin or with backward flow of blood heard by using sound wave blood movement (Doppler study) with standing or during a cough can suggest abnormal **venous function** (**venous insufficiency** or **venous reflux** of blood). Additional feeling of the groin and thigh can often determine how far the backward flow of blood goes down the leg and can locate any enlarged **veins** (**varicose veins**) that lie underneath the skin. Currently, Doppler ultrasound machines in the **vascular lab** are used for complete assessment of all the **veins** in the leg both deep in the muscle and those lying just under the skin for both **reflux** and the location of **varicose veins**. As significant variation can exist between individuals from one leg to the other, **venous Doppler ultrasound** is invaluable to identify the pathway of drainage for **venous reflux** in directing appropriate surgical treatment. Additional tests of **venous disease** can provide more information about the extent of **reflux**, however direct **venous imaging** with **Doppler ultrasound** provides both anatomic detail as well as direction of blood flow.

Management

The treatment of **saphenous vein reflux** and **varicose veins** depends on many factors. Mild and moderate symptoms can be successfully treated with the use of **compression stocking** therapy (stockings that compress the leg because of their elastic design). Best results are obtained when the stockings are worn while the patient is standing or sitting during the day. Unfortunately, how faithfully the patient uses the stocking (compliance) is made less likely by the inconvenience of wearing the stockings as well as the vanity of using the stockings on a regular basis. **Compression stocking** therapy may also be impractical for those patients in which fashion is a major concern. For these reasons, many patients fail to use their stockings in any standard way. Treatment options for stocking failures or noncompliance thus may prompt more invasive treatments for **varicose veins** and **CVI**. There are four treatments that may be chosen.

Vein Stripping

The traditional surgical treatment for **venous insufficiency** of the **saphenous vein** involves two primary goals. The first is to eliminate backward blood flow (**reflux**) in the **saphenous vein** and its tributaries. The second is to remove unsightly and protruding **varicose veins** through tiny incisions (**phlebectomy**). Improvement to the method over the years has allowed most procedures to be done on an outpatient basis. **Saphenous vein stripping** involves making a small incision at the level of the groin to expose the beginning of the **saphenous vein** and its branches, and a separate incision at or near the level of the knee. A **wire stripper** is then inserted into the **vein** and the **vein** is disconnected at these two incisions. The **vein** is attached to the **wire stripper** and is pulled (ripped) from where it lies. Thus the term "**stripping**" of the **vein** was what it came to be called. Small separate skin cuts (incisions) over the areas of abnormal **vein** dilation are made along the thigh or calf for removal of branch **varicose veins** that were not attached to the main trunk of the **saphenous vein** or where not removed at the time of stripping. **Compression** bandaging is used from the ankle to the level of the thigh for several days to reduce the amount of bruising and discomfort from the procedure. The patient is allowed to walk to comfort levels immediately after surgery with instructions for leg elevation when discomfort develops and when retiring to bed. This procedure can be office based using local anesthesia but more normally has been performed in an outpatient setting with either a general or spinal anesthetic. Recovery is fairly prompt with return to normal function within ten to fourteen days.

Vein Ablation

Three additional treatment regimens have become popular within the last ten to fifteen years as a replacement for traditional **vein stripping**. The use of a laser machine or a **radiofrequency** catheter can each be used to create heat which then clots and eventually scars the **saphenous vein** from within. Each device is inserted into the **saphenous vein** at or near the level of the knee by a small needle puncture of the vein guided by **ultrasound**. A wire is passed up the vein to the level of the groin followed by a special

wire like device which will deliver the heat. Using sound wave pictures (**ultrasound** imaging) the correct position of the treatment catheter is confirmed. A generous amount of dilute numbing medicine (anesthetic) solution is then injected by needle around the area of the saphenous vein from the thigh to the level of the knee. This provides for comfort relief and serves as an insulator so that surrounding fat and skin are not burned. The **laser** or the **radiofrequency** probe is then activated. The heat generated produces a clotting and scarring of the **saphenous vein** from the level of the groin to the knee as the catheter is pulled back and eventually out. The heat destroys the inner lining of the **vein** (**endothelium**) and causes scarring of the **vein** lining and thus closes the **vein** so blood no longer flows. Advantages include few skin cuts, the ability to do the procedure in an outpatient or office setting. There is no need for major anesthetic and there is a more rapid return to normal activity level.

Using a needle to push a drug mixed with air (**foam sclerotherapy**) into the vein which can rapidly irritate, burn and scar the inside of the **saphenous vein** without cutting the skin is another way of stopping backward flow in the **vein**. **Ultrasound** again is used to direct where the drug goes in the **vein**. The drug (**sclerosant**) used is a detergent that is very damaging to the inside of the **vein**. When mixed vigorously with air, the **sclerosing agent** becomes frothy or foam like. This solution is then injected directly into the **vein** and floats into **vein** branches as well. The degree of spread is dependant on the amount (volume) of **foam sclerotherapy** drug used. Care must be taken to avoid foam injection into the deeper **veins** of the leg. This technique is also an office based procedure, well accepted by patients, and can be easily repeated if necessary.

Complications

Complications (problems) occurring **after surgery** are usually relatively minor. These include nonhealing of any cuts to the skin (incisional wound breakdown), infection, or clots occurring in the **deep veins** (development of **deep venous thrombosis**) of the leg, loss of feeling (sensory loss) of skin areas from nerves lying close to the **veins** that may be injured. Recurrence of the symptoms of **CVI** or of **varicose veins** has happened when patients have been followed for long enough (more than 10 years). In these clinical studies, recurrence has been reported to occur in up to twenty or thirty percent of people treated. Many things can lead to a return of symptoms such as the method of treatment, advancement of the disease, and the development of entire new problems. Success, however, is seen in most patients with recurrence of symptoms happening slowly over ten to twenty years.

Problems seen after **vein ablative surgery** include burns, nerve damage and **deep vein thrombosis**. An extremely low but rare complication of stroke has occurred with **foam sclerotherapy** injections in some individuals having congenital heart defects. Nonetheless, **vein ablative techniques** have become popular because the treatment is less invasive and can be less painful with a more rapid return to a full level of activity. The recurrence of **CVI** after **vein ablation** has not been as well studied as **vein stripping**. These techniques have been successful in allowing **vein** care to be delivered in an office based or non-hospital setting.

Conclusions

The superficial leg **veins** can allow the backward flow of blood down the **veins** and result in lower leg swelling, **skin changes** and even skin breakdown (**ulcers**). By removing these non-working **veins**, the symptoms are made better. One can remove these **veins** from the body by pulling them out (**stripping**) or by burning the inside so that they scar shut (**laser** or **radiowave ablation**, or **sclerotherapy**). Other **varicose veins** are usually removed at the same time in similar ways. The patient must know that there can be problems with each method of removal and must believe that the benefits are better than the risks involved.

Commonly asked questions

I have varicose veins. Do I need these to be treated?

Varicose veins are common and many individuals do not experience pain or discomfort with them. While cosmetically unsightly, no treatment is necessary if no other symptoms are present. **Varicose veins** can develop **blood clots** that produce pain, redness and tenderness to the touch and these symptoms can last two to four weeks. This is called '**superficial thrombophlebitis**.' It is not a major problem in most cases and will get better with minor or no specific care. Other symptoms seen with **varicose veins** are pain with standing, itching, skin irritation, leg swelling and other less common symptoms. Such symptoms can lead to the decision to treat. Otherwise, the decision to treat **varicose veins** with out symptoms is best addressed between the patient and the physician on an individual basis.

Are stockings necessary to wear for CVI or varicose veins?

CVI is responsible for a reverse direction of flow that allows blood pooling within the **veins** of the leg at the level of the ankle. An analogy would be a waterfall with the top of the falls being at the level of the groin and the bottom of the pool near the ankle. The larger the waterfall (more significant **reflux**) creates a greater amount of pooling at the ankle. With mild to moderate symptoms, **compression stocking** therapy is extremely effective at controlling leg swelling. While it does nothing to treat the cause of the disorder, stockings can control the symptoms quite well. Stockings are only as good as how faithful the people are about wearing them. The decision to use a "below the knee" stocking versus a "thigh high" stocking is at the discretion of the physician and patient given the extent of the **venous insufficiency** involved.

Which is better, vein stripping or "laser" surgery?

This decision is best made with your doctor once after a careful history of **vein** problems and an examination has been completely. The doctor will also have a **venous Doppler ultrasound** study of all the **veins** in your leg to allow for a better impression of your

special **venous disorder**. If additional incisions are needed for removal of large numbers of **varicose veins** or the **saphenous vein** is extremely large, **vein stripping** may be in order. Otherwise the less invasive **vein ablative treatments** may well be possible and give an equally good outcome.

Are varicose veins hereditary?

Varicose veins do run in families. The appearance of **varicose veins** also increases with the number of pregnancies a woman may have during child bearing years.

Figure 1: An artist's depiction of lower leg varicose veins in the calf demonstrating the ropy like characteristics of varicosities beneath skin.

CHAPTER 17
SURGICAL THERAPY FOR DEEP VALVE INCOMPETENCE

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Introduction

Deep vein valvular incompetence happens when the **valves** in the **veins** (tubes that deliver the blood from your leg back to your heart) of your leg stop working well allowing blood to run backward into the leg after it has been pushed forward. These **veins** run along side the major arteries (blood vessels that bring the blood from your heart to the legs) and both travel deep within the muscles of the leg. The **veins** split below the knee into the three paired **tibial veins** of the calf. Within the **veins** are **valves** at the level of the groin, near the middle of the thigh, behind the knee and in the smaller **veins** in the calf. When working well, **venous blood flow** travels in one direction towards the heart pushed forward by the muscle in the foot, calf and thigh. **Valvular reflux** occurs when **valves** quit working and allows blood to flow in the reverse direction. **Venous insufficiency** can cause a number of problems from leg swelling to **skin changes**, including **ulcers**. Venous **valves** are made of two thin leaflets lying within the leg **vein** which meet in the middle of the vessel for proper closing. The **valves** are similar in structure to a heart valve although on a much smaller and thinner scale. Generally, **deep vein valve surgery** is done only for those people in whom **compression stocking** therapy and removing the problems of the **superficial veins** (**saphenous vein ablation**) have failed to take care of symptoms. These people usually have **skin changes** and **ulceration** associated with the **venous incompetence**.

Diagnosis

A good history can help your doctor to know if the **reflux** and **valvular incompetence** is due to primary disease, which happens because the vein itself enlarges resulting in the **valve** leaflets not being able to meet or from **venous thrombosis**, which means the **valve** itself was damaged by **blood clotting** and scarring. Approximately one-half of the patients will be found to have either **primary disease** or **post-phlebitic valve damage**. A good physical examination shows the effect the **venous incompetence** is having on your leg: **varicose veins** are present, swelling is present, **skin changes** have occurred or an **ulcer** is present. A very detailed **ultrasound** study gives a road map of the entire anatomy of the leg veins. Swelling of the leg with standing and during walking using an air boot (**air plethysmography**) to measure the changes can give the doctor data of leg swelling and **venous reflux**. An evaluation of any **blood clotting** disorders is also useful to determine if previous **venous thrombosis** might be a problem during and shortly as surgery.

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Surgical Options

In general, there are three ways to fix vein **valve reflux**. The goal of each method is to put a working **valve** back into lower leg **vein** system and by so doing to prevent further **reflux**. The method used depends on what your surgeon believes is best as well as the location in the leg of **valve incompetence** and whether the **valve** leaflets are damaged or not. Several studies have shown that fixing or placing a working **valve** in the **femoral** (or groin) location works well **(Figure 1)**. Repair of the **popliteal vein valve** which is located behind the knee can be a second option. First described by Dr. Kistner in 1968, directly fixing the **valve** is very successful and lasts a long time. **Venous valve repair** requires magnification to do the best job and it is a very demanding work which must be done perfectly. The **direct valve repair (Figure 1)** of Dr. Kistner requires opening the **vein** to allow the surgeon to look at the **valve** leaflets and then to place sutures to "cinch" or tighten up the **valve**. Once this is done, the **vein** is closed so blood can flow normally again. This tightening of the **valve** parts allows for proper closing. If one is familiar with sailing, it is much like the cinching of a sail to allow it to catch more wind. Fine filament sutures smaller than a human hair are used to retack or cinch the valve to the correct tightness. A simple test done by pushing blood from below to above the **valve** while still holding any more blood form coming from the leg and seeing if the **valve** now works (the "strip test") shows that the repair is working well. The patient is given **blood thinners** (**heparin**) during the operation to make sure no **blood clots** occur and is continued during the short hospital stay while changing to **blood thinner** (**warfarin**) that can be taken by mouth which is continued for eight to twelve weeks. Those patients having prior venous damage from **venous thrombosis** (**blood clotting**) may need longer term **anticoagulant** (**blood thinning**) treatment.

Other ways to place a good **valve** into the refluxing lower leg **vein** system may also work. One can cut the main **vein** in the **incompetent veins** and suture it into place below one of the other **veins** in the lower leg that has a working **valve** (this is called a **valve transposition** and involves a **vein relocation**) **(Figure 1)**. There may not be such a **valve** present in the lower leg making this approach impossible and there is some concern that overtime the extra work this **valve** must do might cause the **vein** to dilate causing this valve to also fail.

Axillary vein valve transfer (Figure 1) originally described by Dr. Raju in 1981 is used when **direct vein valve repair** or **vein relocation** is not possible. The **axillary veins** near the armpit are of similar size to the **femoral veins** in the thigh. A segment of **vein** with a good functioning **valve** is taken from the arm **veins** through a small incision in the armpit. This **valve** segment is then placed into the lower leg **incompetent vein** system by suturing it to both ends of the cut deep leg **vein**. Occasionally a plastic cover is placed over the **valve repair** site to prevent late **vein** dilation.

Complications

Complications or problems occurring during the operative experience involve approximately ten percent of patients. These are most commonly hematomas or bleeding in the area of operation or collection in the wound of other bodily fluids. A re-operation to drain these fluids may be needed to make sure the **valve** continues to work well. **Thrombosis (clotting)** of the **valve repair** site occurs in roughly five percent of patients despite **anticoagulant** treatment.

Results

Improvement in symptoms including stopping pain and swelling can be found in sixty to eighty percent of patients who have **venous reflux** due to **primary valve dysfunction**. Most patients are able to stop or limit stocking use after successful operation. The results are not as good for those people who have **valve** surgery because of prior **vein thrombosis** and extensive **post-phlebitic** (scarring) **changes**. Nonetheless, two-thirds of patients can be found to have complete **ulcer** healing at twelve years following successful surgery. Best outcomes can be seen in those centers which have the surgeons, tools and skills needed for these demanding operations available.

Conclusions

Vein valves that do not work will cause blood to flow backward in the **veins** into the legs. This leads to problems with swelling, **skin changes** and even breakdown of the skin (**ulcers**). There are ways to stop this abnormal backward flow of blood by fixing the **vein valves**. If the **valve** is still present but just not meeting properly, the **valve** can be fixed with fine sutures. If the **valve** is totally damaged, one must place the refluxing system below a working **valve** in another part of the leg **veins** (**transposition**) or must take one from the arm as a transplant. Other techniques are being investigated but so far these are the more common ways to fix the problem.

Commonly asked questions by patients

When such I ask my doctor about deep vein valve surgery?

Not all patients with **valve reflux** and **venous insufficiency** or who have had prior episodes of **venous thrombosis** need **deep vein valve reconstruction**. More commonly done and less invasive methods such as **compression stocking** therapy and treatment of all **superficial vein reflux** is considered before recommending **valve reconstruction**. The majority of patients can be managed with these methods to provide for **ulcer** healing and reduction of leg swelling. If these methods fail, **direct valve surgery** would be considered. Knowing the exact cause of the **venous reflux**, whether it be **primary valve dysfunction** or secondary to **venous clot** damage, is important to know so that the surgeon can give the patients a good idea of the possible success and durability of the procedure. This conversation should occur after the appropriate workup and the diagnosis has been completed.

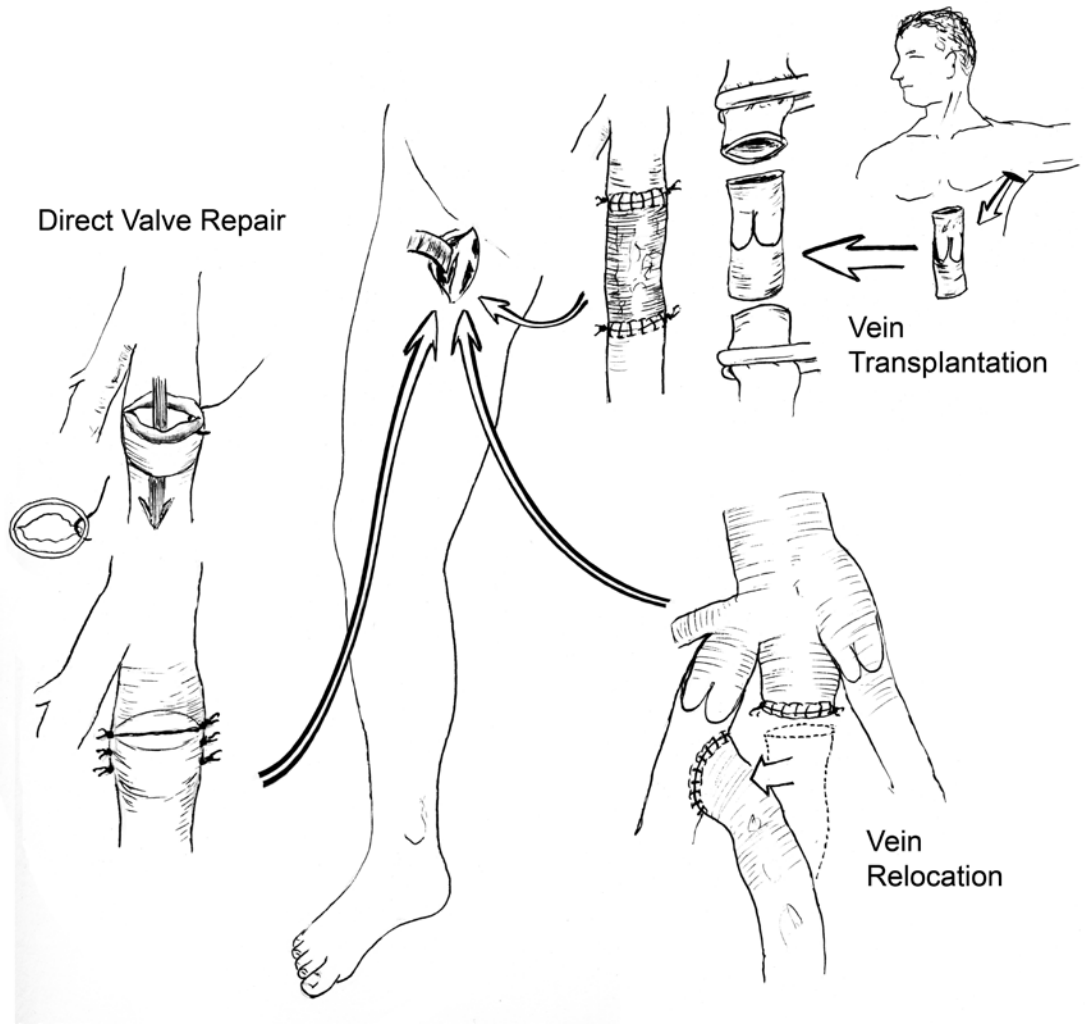
How long will I need to be on Warfarin treatment?

The length of time necessary for chronic **anticoagulation** (**blood thinning** drugs) after **valve repair** is dependent on the surgeon's thoughts, type of repair, and the reason for **valve incompetence** in the first place. **Anticoagulation** for eight to twelve weeks is standard for **direct open vein repair**. Longer duration of therapy may be necessary for those individuals who have a history of prior **clotting disorders**.

What happens to the arm if the vein is taken from that location to be transplanted to the leg veins?

Removal of the **axillary vein** from the arm surprisingly causes little problems in most cases. There are many collaterals (small **veins**) within the arm that allow for continued drainage of blood from the arm without significant swelling or pain. Rarely some arm swelling is seen but is very manageable.

Figure 1: The artist has drawn pictures that show different ways of surgically placing a work valve in the lower leg deep veins to prevent problems with deep venous reflux. The picture of the leg shows a cut in the groin in many of these veins repairs are preformed. The picture to the left of the leg shows a direct repair of a floppy valve using very fine sutures to tighten the valve edges and make it work again (direct valve repair). The picture in the upper right shows taking a working valve from the arm and sewing it into the lower leg deep vein to prevent reflux in that system (vein transplantation). The picture in the bottom right shows placement of the non-working or incompetent major vein below a working valve in another part of the lower leg deep veins (vein relocation or transposition).



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CHAPTER 18
ILIAC AND VENA CAVA OBSTRUCTIVE VENOUS DISEASE THERAPY

Original author: Peter Neglén
Abstracted by Gary W. Lemmon

Introduction

Ilio-femoral (**iliac vein** and upper **femoral vein**) and **vena cava vein thrombosis** involves **blood clot** formation in the large **veins** leading from the leg (groin area) and going into the pelvis (**iliac veins**) and draining into the **vena cava** (the largest **vein** in the abdomen) with goes directly into the heart as it enters the chest. Only twenty percent of clotted **iliac veins** will completely "recannalize" or open back up with **anticoagulant** (**blood thinning drugs**) treatment. Sometimes, severe **compression** of the **iliac vein** can occur when it is repeatedly hit by the artery that crosses over it (the iliac artery). As similar constriction (narrowing) of the **iliac vein** can be seen in a young pregnant lady's with a large uterus with baby within that gradually narrows the **iliac vein** resulting in a **blood clot** (**vein thrombosis**). Although happening more commonly on the left side, over twenty-five percent of the time it can happen to the right **iliac vein**.

While **blood clots** in the upper leg and behind the knee (**femoral** and **popliteal veins**) often cause mild to moderate symptoms and can be managed with **compression stockings**, **thrombosis** or extrinsic **compression** bad enough to produce an **obstruction of the iliac vein** causes more problems due to the larger volume (amount) of blood that is trapped in the lower leg. Recanalization or reopening of the clotted **vein** with **anticoagulant** therapy alone occurs in only about a fourth of patients. Therefore a majority of patients with this problem remain symptomatic with limb swelling, pain on standing, and the other signs of long term **chronic deep venous obstructive disease** including **skin damage** and even **ulcers**.

Diagnosis

The significant leg swelling and pain experienced with **iliofemoral deep venous thrombosis** should result in a workup including **duplex ultrasound** (sound wave imaging of the vein) in the leg, pelvis and abdomen (if possible). This study is not as good in terms of imaging of **veins** significantly above the level of the groin. To see the pelvic and abdomen **veins**, noninvasive studies such as a **CT (computerized tomography) scan** or **MRI (magnetic resonance imaging)** may be needed to find out where the **blood clot** is located. Invasive **contrast venography** (injecting x-ray dye (contrast) right into the correct **vein**) along with **intravascular ultrasound** (a sound wave device place into the **vein** to take pictures from the inside) may be needed to determine with certainty how long the **clot** is and what **veins** are involved **(Figure 1)**. **Intravascular ultrasound** has proven to be more reliable for determining the type of disease, its extent and exact location. Multiple levels of obstruction may occur during an attack of **venous thrombosis**. Extrinsic **compression** (something pushing on the **vein**

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from outside the **vein** itself) is often found to be located at just one spot near where the **iliac veins** come together to make the **vena cava**.

Therapy

Open surgical techniques such as a **surgical bypass** to jump over the obstructive or stenotic **vein** are often not needed with the current use of **venous stenting** (placing a metal support device within the **vein** to help keep it open). **Stenting** of the affected **vein** segment is needed to keep the **vein** open after dilating it up with a balloon **(Figure 1)**. The **stenting** procedure is not very invasive, involves little discomfort, and usually does not have many problems during or after the procedure. The **femoral vein** in the groin or the **popliteal vein** behind the knee are the places where a needle is pushed into the **vein** through the skin after numbing medicines have been given. A wire is then passed up the vein to the place in the pelvis or abdomen where the occluded or narrowed **iliac vein** is located. With some delicate movement of the wire it can often be placed into the obstruction and past into a normal **vein** above the disease. A balloon can then be passed to the area of disease and blown up to its optimal size to increase the size of the **vein**. A stent is then placed in the same area which opens up to the new ballooned size and pushes from the inside to keep the vein open. The stent must cover the entire area of diseased vein or its usefulness will not last long. Long term **anticoagulation** (**blood thinning drug**) is not needed for those patients with only extrinsic **compression** problems. The surgeon may need you to take such medicine if the initial damage was done as a result of **blood clots**. **Stents** generally do very well in keeping the **vein** open and in so doing improves the patient's symptoms of leg swelling and/or helps the **ulcer** to heal. **In-stent stenosis** (fibrotic tissue developing within the stent after it is placed) is low at five percent in those patients with only extrinsic **compression** and but is slightly more common to over ten percent if the obstruction is due to prior **venous thrombosis**.

Conclusion

Iliac vein or **vena cava vein** blockage or narrowing prevents blood flow from getting out of the leg(s) since these are the major exit sites for this blood to get back to the heart. Removing blockages from these **veins** is possible with techniques that allow one to place a **dilating balloon** and **stent** (metallic support device) inside the narrowed vein. In the rare case that this is not successful, there are surgical procedures which can help.

Commonly asked questions by patients

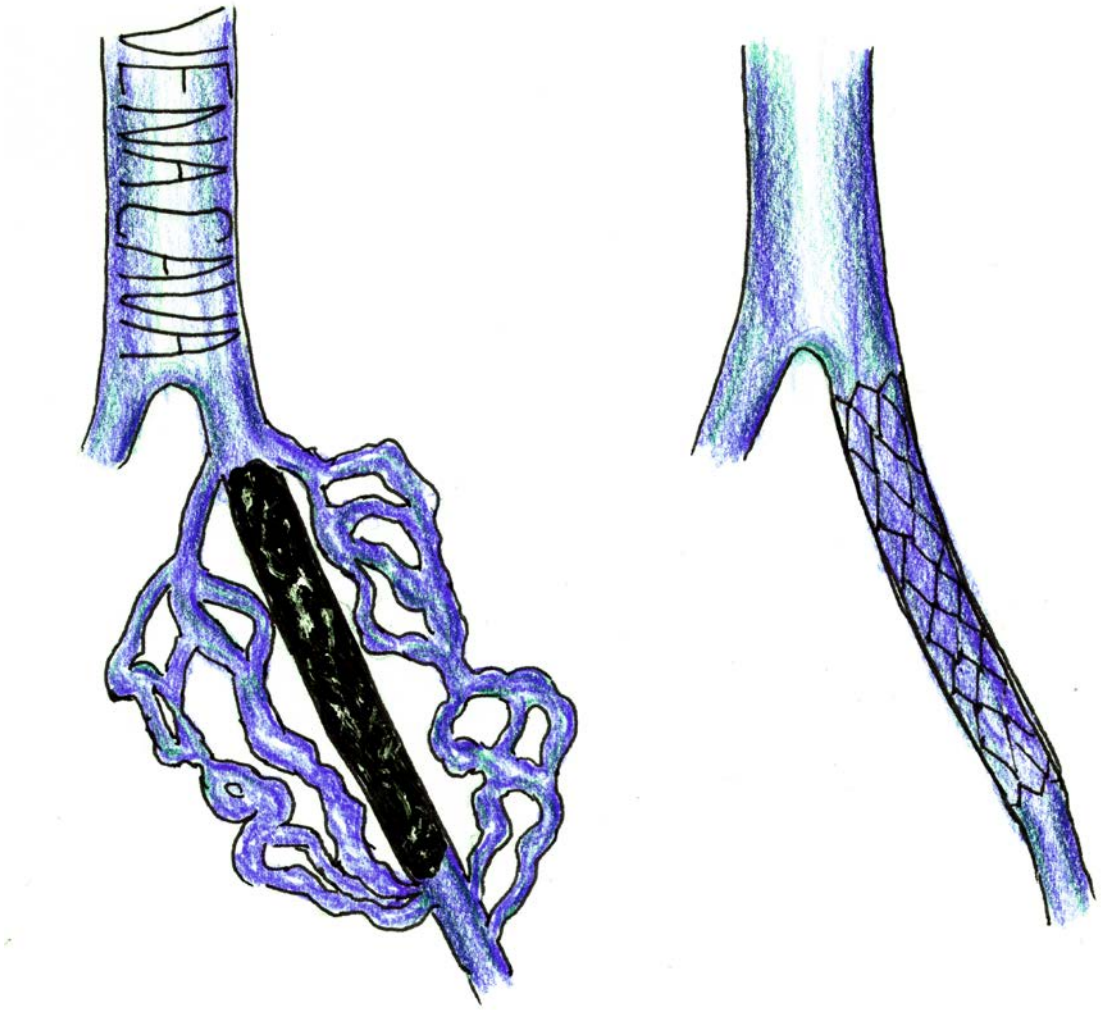
Will treating iliac vein narrowing I have heal my leg ulcer?

Venous disease and leg **ulcer** development has many causes, one of which is proximal **obstruction of the vein** preventing blood from getting out of your leg. This complete obstruction or very tight **vein** narrowing results in to much pressure building up in your leg **veins**. This high pressure in your **vein**s then pushes on the soft muscle, fat and skin especially around the ankle. The result is **skin changes** and then an **ulcer** or open wound. If **iliofemoral** (abdomen, pelvic, proximal groin) **venous thrombosis** or **obstruction** is present with an active leg **ulcer**, **stenting** of those **vein** affected is one way to help heal your leg **ulcer**. Treatment of **venous reflux** (backward flow of blood in your damaged **veins**) in the leg may also be needed before or after the **stenting** procedure. Only your surgeon will know what is the best treatment for your case.

What are the stents made of and can I feel them?

Stents commonly placed in the **iliac vein** are made of one of two metals; either stainless steel or a nickel titanium alloy. The **stents** are "**self expanding**" which means that they open up automatically as they come out of the device (constraining sheath) that gets the stent to the right place in your **vein**. The **stents** are made for large vessel sized (diameters) and open, sometimes with the help of a **balloon dilation**, to go to twelve to fourteen millimeters in diameter (about the size of a quarter). They can be gotten to the **pelvic veins** by placing a needle in a more easily punctured leg **vein** in the groin or behind the knee **vein** (**femoral** or **popliteal vein**) and then pushing the **stent** which is held in a plastic covering on a long wire to the proper place. Once the plastic covering is pulled back and the **stent** opens in the vein, it delivery wire is taken out and gentle pressure of the needle stick sight stops any bleeding. The **venous system** is a low pressure blood containing tube and so there is rarely a bleeding problem. Many of these procedures can be done in an outpatient setting.

Figure 1: The first picture shows an iliac vein occlusion with many collateral veins (smaller veins going around the blockage) present to help decrease the pressure in the lower as a result of the blockage. The second picture shows a metal stent in the iliac veins which has opened it fully so the collateral veins are no lower seen because the blood is going through the larger and more functional iliac vein. The excess vein pressure in the lower leg has been corrected.



CHAPTER 19
VASCULAR MALFORMATIONS

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Abstracted by Raghu Motaganahalli

Introduction

The difference between a **hemangioma** and **congenital vascular malformations (CVMs)**, both of which are commonly called "birth marks", is important to recognize.

Hemangioma

Vascular **hemangiomas** are a big concern to the family and are rather common. A **hemangioma**, commonly known as a **"strawberry birthmark"**, is the most common benign tumor of infancy. They are often not seen right at birth and most are seen in the head and neck but can sometimes involve other parts of the body. They are more common in girls. These vascular growths have a very active **blood vessel** lining so grow very fast during the first year of life to the fear of the parents but then stop growing and actually get smaller very slowly so that most have disappeared by school age. Because the presence of the **hemangioma** can be alarming to the family, education and reassurance that it will go away is important information. In rare cases, since the head and neck is involved, vision problems, feeding problems and breathing problems can be seen. If the **hemangioma** involves the liver, heart or lungs, breathing problems or heart failure can occur. Generally, a drug used to reduce swelling (steroids) will take care of the problem. In very rare cases, **sclerotherapy** (injecting drugs directly into the tumor which can scar the insides) will decrease the size and allow normal bodily functions.

Congenital vascular malformations (venous malformations emphasis)

Overview

CVMs are different in that they have no rapidly growing **blood vessel** lining and do not go away. **CVMs** occur in approximately 1% of the general population. They occur for no apparent reason and rarely run in families (no genetic reason). **CVMs** involve **arteries** (**blood vessels** delivering oxygen and food to your cells from your heart), **veins** (**blood vessels** removing waste from your organs back toward the heart), and/or **lymphatic vessels** (**blood vessels** that clean up any excess in the tissues that the **veins** do not collect). A doctor's meeting held in Hamburg, Germany, came to an agreement that **congenital vascular malformations** should be separated into five different groupings, namely 1) predominantly involving **arterial defects**; 2) predominantly **venous defects**; 3) combined **artery** and **vein defects** (shunting of blood between the two) 4) predominantly **lymphatic defects**; 5) combined **vascular** (all the above in addition to **capillaries** which are the normal connections between **arteries** and **veins**) **defects**. Each

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is further divided into when the normal development of normal **blood vessels** were stopped (arrested) during pregnancy. If stopped early on the way to forming the normal **arteries** and **veins**, the incompletely formed **blood vessels** have some of the growth properties of primitive **blood vessels** which means that they can grow when influenced by puberty, pregnancy or surgery. The lesion seen can be very small and well defined or more extensive involving muscle and even bone. In contrast, the more mature form does not grow, has main channels formed but they are formed abnormally leading to a total lack of normal **vessels** (aplasia), blockage where there should be normal **blood vessels** or the **blood vessels** can be too big (dilation). It is important to separate the different forms since the doctor must treat them differently. **Venous malformations (VM)** account for nearly 50% of all **CVMs** and there are a few more ladies than men affected.

VMs which stop maturing later in pregnancy

VMs which stop maturing later in pregnancy are present at birth but often **symptoms** are not noticed until the late teens and are most often seen in the lower leg. **Symptoms** (pain, swelling, skin changes and even skin wounds called ulcers) common to all **venous disorders** brings the patient to the doctor. The patient may have early **varicose veins** or locally enlarged **veins** (**phlebectasia**). There can be blockage of **deep veins** either completely (**aplasia**) or partially (**hypoplasia**). **Aplasia / hypoplasia** of the **deep veins** may require the **superficial veins** as the exit route of blood from the legs or arms. Removing the **superficial veins** in these patients could cause major problems. Commonly, these patients develop a "**marginal vein**" on the outside of the leg which is not usually present and may be very important to getting blood out of the leg. Narrowing of the left **common iliac vein** (the big vein in the pelvic and abdomen which allows blood to exit the left leg) can happen as the right **common iliac artery** crosses over it to get to the right leg. It is called the **May-Thurner syndrome** or **iliac compression syndrome**. Compression and repeated hitting of the left **common iliac vein** from the right common iliac artery as it hits the underlying spine damages the **vein**. The majority of children affected are teenage girls on oral contraceptives showing up with a swollen left leg. If **blood clots** form because of the narrowing and injury, it is called **deep venous thrombosis**. The **deep veins** can be too large. **Avalvulia** or the lack of **vein valves** in both the **deep** (in the deep muscles) and **superficial** (just under the skin) **veins** often runs in families (has a genetic cause) seen as swelling and **varicose veins** when the child starts to walk. Significant **venous reflux** (backwards flow of blood in the **veins** into the lower leg) and sustained high pressure in the **veins** leads to swelling and **varicose veins** often before puberty. There can be very focal dilations called **aneurysms** which can become filled with **blood clot** (**deep venous thrombosis**). These local dilations can happen in the legs, **veins** in the abdomen or even in the chest. Early studies to show that a **VM** is the problem should be a **magnetic resonance imaging** (**MRI**, a study using magnets to see into the body). A **venous duplex** study (using sound waves to see into the body) will aid in determining what **veins** are involved and how the body is being affected. **Venography** (an X-ray study using drugs injected into the **vein** which help to see the **veins**) may be used to clearly show **vein** connections and prevent incorrect treatment during procedures. In general, these tests tell what **veins** are affected, if they are working well and what can be done to help **vein** blood flow. If acute **blood clots** are the problem, then ways to

remove the **blood clot** (clot busting drugs) or drugs used to thin the blood and stop more clot from forming (**anticoagulants**) are the correct treatment. If swelling in the only problem, **compression stockings** (external force pushing from the skin) can sometimes take away the pain, decrease and even control the swelling. If the stockings are not working and only the **superficial veins** are allowing backward flow of blood resulting in **varicose veins**, there are many ways to eliminate the **vein** including removing it by pulling it out (**stripping**) or more recently by scarring the inside of the **vein** with heat (**lasers** or **radiowaves**) or damaging drugs (**sclerotherapy**). The latter is usually done only when the child is somewhat older. If **deep veins** are involved, **superficial veins** can only be removed if the **deep veins** are open to allow blood to get out of the leg. If the **deep veins** are blocked, dilating the **vein** (**percutaneous venoplasty**) and placing a device in the vein to keep it open (a **stent**) can sometimes correct the problem. If the **deep veins** have no **valves** and **compression** is not working, then sometimes the surgeon can fix or replace the **valve** but this is uncommonly needed. Whether simple **vein** dilation should be removed or not is a decision your doctor must make with the patient's thoughts taken into consideration and that decision is determined by where the dilation is and what other structures lay close.

VMs which stop maturing early in pregnancy

Those **VMs** with growth that has stopped early in pregnancy do not look much like normal **veins** but more like a bundle of soft, spongy blood lakes that can flow into and become part of muscle, bone, fat and other organs. Since they go into fat, muscle, bone and other organs; it is very important the diagnostic studies are used which can picture where the abnormality is located, how the **veins** meet up and where, and what other organs are involved so a proper plan for treatment can be made. This usually means that **MRI** and invasive studies (studies using needles place into the body) which directly put drugs easily seen by X-ray into the **veins** (**venogram**) and sometimes arteries (**angiogram**). The desire is to get rid of the malformations with as little damage to surrounding body parts. When causing symptoms from swelling and when compression is not working, the usual way to get rid of the abnormal cluster of **veins** is to put drugs that damage the **VM** directly into the **veins**. This means placing a needle right into the **VM** or by placing a needle into the **vein** which is far away and directly a very small plastic tube (a catheter) into the problem area. When properly in place; a drug (**sclerosant**), which can damage the **VM**, is pushed into it. The **sclerosant** can be high grade alcohol or a number of other drugs. Unusual problems sometimes seen after injecting these drugs are open areas and infection in the skin, **blood clots** within the **veins**, and even **blood clots** moving to the lungs (**pulmonary embolism**) that can stop you from breathing causing death or even strokes. Rarely is open surgery useful and in many cases simple compression and elevation is the best treatment.

Complex Venous Malformations

Venous malformations that have other **congential vascular malformations** (complex **VMs**) as a part of the overall syndrome are rare but of concern if a person in the family has the problem. Although we are trying to move away from naming particular disorders,

many people have come to know special conditions. **Complex venous malformations** are divided into those without (**Maffucci Syndrome, Proteus Syndrome, Klippel-Trenaunay syndrome**) and those with direct artery to vein connections (**arteriovenous shunting**) (**Parkes-Weber Syndrome**).

Maffucci syndrome

The **Maffucci syndrome** has a **venous malformation** (often large and in the fatty tissue just under the skin (subcutaneous)), abnormal growth of cartilage (a type of soft bony part of the body), and bone disfigurement. About 20% of patients can have changes into cancer which means that careful following of the patient is needed to catch any such changes early. If this does happen, removing the lesion is needed since other therapies do not work.

Proteus syndrome

The **Proteus syndrome** has several **blood vessel** malformations (**capillary, venous** and **lymphatic malformations**) combined with abnormal growth of bones, muscles, and fatty tissues. Children are usually born without deformity. As they age; tumors, skin, and bony growths appear, get worse and may involve more than half the body. Connective tissue nevi (birth marks) on the abdomen, hands, or nose are telltale signs of the disease. **Venous malformations** play a big role in the risk for **blood clot** in the **deep veins** (**DVT**), **blood clots** traveling to the lungs (**pulmonary emboli**), and premature death. No cure exists.

Klippel-Trenaunay syndrome

The **Klippel-Trenaunay syndrome** is a mixed **capillary, venous,** and **lymphatic malformation**. Symptoms vary from mild **varicosities** to massively enlarged unilateral lower limb involvement. A skin blemish or hyperpigmented area (dark birth mark on the skin) is commonly seen. X-ray studies may find lower leg **deep vein** abnormalities like to small or absent **veins** in the leg or abdomen and the presence of a **vein** which should have been replaced during the pregnancy (persistent **sciatic vein**). Mild **varicosities** and symptoms are managed best with **compression stocking** and leg elevation. If a particular skin malformation bleeds often then **sclerotherapy** might be a good treatment. Although recurrence of **venous varicosities** after removing the **veins** (**open, laser, radiowave removal**) is common, such treatment can help to decrease local pain, swelling and improve cosmetic image if the deep system is good enough to remove blood from the lower leg.

Parkes Weber syndrome

The **Parkes Weber syndrome** is distinguished by ***high-flow*** (rapid blood flow in the abnormal **blood vessels**) **arteriovenous fistulas** and problems that occur with such connections. Brightly stained skin which is warm to the touch as well as certain noises (bruit and thrills) which your doctor listens for can make the doctor think of this problem.

Problem with heart failure usually happen later in life but if the connections are very large this can occur even in infants. Most of these direct **artery** to **vein** connections are not easily gotten to for surgical removal. Thus, **sclerotherapy** has become the way to control symptoms.

Conclusions

A **hemangioma**, commonly known as a **"strawberry birthmark"**, is the most common benign tumor of infancy. These vascular tumors grow very fast during the first year of life to the fear of the parents but then stop growing and actually get smaller very slowly during childhood so that they are usually gone by school age. Only in rare cases is any treatment needed. **Congenital vascular malformations** are the result of **blood vessels** not maturing (going to full development) while the infant is still in the mother's womb. If this happens early, the abnormal **blood vessels** do not have the form usually seen with **blood vessels** and appears more like a spongy mass which can involve neighboring body parts. If this happens later in the pregnancy, the **blood vessels** look more normal but are abnormally small, abnormally large or have unusually connections with other **blood vessels**. How bad the symptoms are will determine the need for treatment. As a part of **congenital vascular malformations**, the **venous malformation** is the most common and possibly the easiest to manage. **Congenital vascular malformations** do not go away and will require a lifetime of care.

Commonly asked Questions

My child has a reddish spongy mass on the cheek, what should I do?

You should see your pediatrician. He can tell if this is a **hemangioma** or what some people call a **"strawberry birthmark"** which may look bad now but will go away with time. This is a rather common problem in children. There are other much less common problems which have a similar appearance but must be managed differently. One such condition is the **congenital vascular malformation**, which results from an abnormal maturing of **blood vessels** as the baby develops the mother' womb. The doctor will know how to tell the difference and when more testing or treatment is needed.

My doctor thinks that my baby had a congenital venous malformation, what does this mean?

A **congenital venous malformation** means that some of the **blood vessels** which should have matured into normal **veins** did not make it to the final stage of growth. If this happens early, the abnormal **blood vessels** do not have the form usually seen with **blood vessels** and appears more like a spongy mass which can involve neighboring body parts. If this happens later in the pregnancy, the **blood vessels** look more normal but are abnormally small, abnormally large or have unusually connections with other **blood vessels**. How bad the symptoms related to the malformation are will determine the need for further study and/or treatment.

My seven year old has a birth mark on the left leg and back, a varicose vein that I just noticed that goes down the outside of the leg, and his leg on that side may be a little bigger than the other side. What could be the problem?

Your child might have a syndrome called the **Klippel-Trenaunay syndrome** which involves an abnormal maturing of **veins, lymph blood vessels** and the normal connection between **arteries** and **veins** (the **capillary blood vessels**). It is a **congenital vascular malformation** which has been present since birth but just noticed now because your child is up walking and playing. X-ray studies may find lower leg **deep vein** abnormalities like too small or an absent **vein** in the leg or abdomen and/or the presence of a **vein** which should have been replaced during the pregnancy but remains to help get blood out of the leg. The lateral **varicose vein** my also be helping to remove blood from the leg so is important to have. Early on all that is needed to take care of symptoms is the use of support stockings. Your doctor will be able to tell you if this is the problem and, if so, if other treatment will be needed.

My child has a congenital venous malformation, are we at risk for other children with the same problem?

Most **congenital venous malformations** do not run in the family, in other words, are not genetically determined. There are some that do have a genetic basis especially certain syndromes. You would have to discuss this with your doctor to know for sure if this is a concern for your family.

CHAPTER 20
LYMPHEDEMA DIAGNOSIS AND THERAPY

Original authors: Audrea Cheville, Cindy Felty, Gail L. Gamble, Peter Gloviczki, Thom W. Rooke, David Strick
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Introduction

Lymphedema is a condition in which an excessive amount of **lymph** (liquid with some solid parts) collects in one place in the body, causing **edema** (swelling) and later if not properly treated, skin problems. **Lymphedema** occurs most frequently in the arms and legs. **Lymphedema** is most often a chronic condition and rarely cured, but it can generally be controlled by carefully following a treatment program.

Lymphatic System

The **lymphatic system** plays a very important part in keeping your body healthy. It works as part of your body's cleaning system, by collecting and removing **lymph** fluid (which contains proteins, fatty acids, waste from cell feeding and bacteria that enter the body) from the body. **Lymph** fluid is taken up in **lymphatic vessels** and passes through **lymph nodes** where harmful substances are destroyed and cleaned by special infection-fighting cells that are present in our body (by the **immune system**). After **lymph nodes** have filtered the **lymph** going through them, the **lymph** fluid returns to join the **blood** pumping through your **veins**. When this protein-rich fluid is not removed because of damage to the **lymphatic vessels** themselves or the **lymph nodes**, the result is **lymphedema** or swelling of the leg or arm. If the **lymph vessels** do not develop normally, or if the **lymph nodes** or **lymph vessels** have been removed or damaged (such as with radiation from cancer treatment, or from repeated infections in the leg or arm), **lymph** fluid may collect (stay) in the soft tissues of the affected body part causing **lymphedema**.

Types of Lymphedema

There are 2 types of lymphedema: **primary lymphedema** and **secondary lymphedema**. **Primary Lymphedema** occurs when the development of **lymph vessels** and/or **lymph nodes** is abnormal. This condition may be present at birth, but may only show up as **edema** at the time of puberty or even later in life. **Primary lymphedema** is much more common in women and occurs most often in the legs. A significant number of patients with **primary lymphedema** (perhaps as many as 10%) are thought to have chronic **reflux** (back flow of lymph fluid in the **lymph vessels**). This includes patients with congenital **lymphatic hyperplasia**, which in its most extreme form is sometimes known as **"megalymphatic" disease**. There are 3 types of **primary lymphedema**. **Congenital lymphedema** presents at birth and accounts for about 10% of all cases. **Lymphedema praecox** shows up between the ages of 2 and 25 years old and represents about 80% of

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all **primary lymphedema**. Finally, **lymphedema tarda** is generally noted first after the age of 35 and accounts for 10% of these cases.

Secondary Lymphedema has 2 causes. The first is obstruction or blockage of **lymph** fluid movement in the **lymph vessels** or **nodes**. The cause can be injury, infection, blockage from tumor invasion, radiation therapy, or by surgical removal or damage of the **lymph nodes** or **lymph vessels**. Surgical damage of the **lymph system** may be the most common cause in the United States. Scarring of the **lymph vessels** or **nodes** is the final result of most causes. The most common cause of **lymphedema** world wide is **filarial disease**, caused by infecting worms. These are parasites (worm type invaders) that get into the patient and damage the **lymph vessels** as well as causing other problems. The world health organization estimates that the number of people affected worldwide by this problem may approach 100 million. A less common cause of **lymphedema** is overproduction of interstitial / **lymphatic fluid**, the second cause of **secondary lymphedema**. High body temperature, injury or inflammation (infection) are thought to produce **lymphedema** by this mechanism. Most of these conditions resolve in time and so does the **lymphedema**. Chronic overload occurs when the **lymphatic system** is repeatedly damaged over time due to recurrent infections, chronic **vein** problems, or obesity. These types of overload to the **lymphatic system** can cause **lymphedema** to progress more quickly.

Symptoms of Lymphedema

There are four **stages of lymphedema**. In the **first stage**, the body can make up for any decreased movement of **lymph fluid** so no physical changes are seen (**latency stage**). Stage 2 is called "**reversible lymphedema**" and even though swelling is seen no actual damage has occurred to the body and so the swelling can be removed by medical means. Stage 3 (called "**spontaneously irreversible lymphedema**") means that actual damage (scarring or fibrosis) has occurred to the body and the only way to change the situation is by surgery. The final stage is called "**lymphostatic elephantias**", or elephant like appearace, with severe thickening and hardening of the tissues to the point of disfigurement. What the doctor and patient sees with **lymphedema** is swelling in the leg, arm or other body part, limited movement, feeling of heaviness, tightness or aching, recurrent infection, and hardening and/or thickening of the skin. **Lymphedema** does not cause focal pain (pain in a localized, specific area). You should contact your doctor if you are having pain, weakness, or numbness in the affected body part.

Diagnosis of Lymphedema

The **diagnosis of lymphedema** is based on a history of progressive and non-painful swelling especially into the foot or hand if the limbs are affected. The physical findings are swelling in the leg, arm or other body part, limited movement, feeling of heaviness, tightness or aching, recurrent infection, and hardening and/or thickening of the skin. An **ultrasound study** uses sounds waves to bounce off underlying structures to make sure that the **edema** that is present is not as a result of a new **blood clot** or other damage to the **vein**. **Magnetic resonance imaging (MRI)** is a study using magnets to see into the body

and can help to prove that the type of tissue involved is in the fatty tissue under the skin and is not a **vein** related problem. **Lymphoscintigraphy** is the most common test use to show that the **lymphatic system** is the problem. The study involves giving an injection of a small amount of **technetium 99m-labeled colloid** (a substance that can be seen by this type of study) into the space between the toes and takes pictures of the leg or arm or other affected part of the body as the colloid is taken into then **lymph system**. This test is very sensitive for diagnosing **lymphedema**. The **lymph system** is followed for up to 24 hours or until the medication is taken up in the liver. The test shows if there are areas where the medication is not taken up indicating obstruction or damage to the **lymph vessel** system. **Lymphangiography** is not used in most cases since it can cause **lymphedema** but in certain cases it is the only test that can actually show the anatomy of the **lymph vessels**. It is usually done on patients with **chylous reflux** or abdominal or **thoracic chylous fistulas** or connections in the abdominal or chest cavity that makes **lymph fluid** come in contact with parts of the body that it usually does not connect with. If actually surgery on the **lymphatic system** is planned (microsurgery), a **lymphangiogram** may be required.

Medical Treatment of Lymphedema

Early treatment is key in achieving long-term control of **lymphedema**. The medical treatment of **lymphedema** involves two phases: **reduction therapy** which focuses on actively decreasing the swelling and maintenance therapy which is used to maintain or keep the reduced size gained in the reduction phase. **Reduction therapy** involves aggressive wrapping of the limb with **compression bandages** sometime of a stiff material or sometimes with elastic material or a combination of both. **Short stretch or low stretch (not elastic) wraps** are most often used since the **compression** is more constant especially when the patient walks. **Short stretch wraps** have the added benefit of being able to mold most closely to the shape of the affected body part so all of the swelling can be reduced. **Multilayered techniques** are the most popular used. In addition, specially created padding using cut pieces of foam can be applied to specific areas that might have more thickened skin to help remove the protein rich tissue that has accumulated. The idea is to push the fluid out of the affected part by applying the wraps during the entire day with rewrapping whenever the **compression** seems less tight. By measuring the size of the limb frequently during this phase of treatment, the limb can be decreased to its near normal size when compared to the other limb (leg or arm). By compressing the limb the fluid is forced back up into the trunk where it can be reabsorbed by the body. The wraps should only be taken off just before bathing or just before a **massage therapy** session. Exercise is important during **compression** treatment to help get the fluid out of the affected part of the body. A therapist will give the patient special instructions on best exercise. Skin care is also an important part of the care of patients with **lymphedema**. Pressure and friction from the wraps and the appearance of dry skin as the swelling decreases may contribute to problems. The patient must pay close attention to dry or cracked heels, toes or fingers and if prone to athletes foot, the use of an anti-fungal powder or cream to skin creases such as between toes, and skin folds daily or more often may help to decrease infections. A daily moisturizer to prevent dry skin may also help. If you notice a scratch or break in the skin of the affected limb, ask your doctor if an antibiotic ointment applied to the skin might be helpful.

In addition, there are many **lymphedema** treatment programs available in the US. The therapists are specially trained to use a special type of **massage** to decrease lymphedema swelling. This treatment requires certification after special training. This technique called **manual lymph drainage (MLD)** is a type of specialized **massage** which is different from **deep congestive massage** for reducing **lymphedema**. The technique encourages the natural flow of **lymph** fluid out of the affected limb by first clearing out the **lymph vessels** further up the limb before encouraging the **lymph** fluid at the most distal part of the limb. The therapist uses a specific set of hand movements and sequences to gently move the fluid away to more healthy **lymph vessels**. Stimulating (rubbing) the **lymph vessels** allows them to function more normally and filter and remove the excess fluid. Areas of skin may be especially thick with (protein) fibrinous tissue. The **MLD** technique can soften these areas to become more like normal skin. You or a family member may be taught to use this technique at home although it can be challenging to do it on yourself. **MLD** is not used as a therapy alone. It is most often combined with other therapies including **compression wrapping**, exercise, garments, and skin care.

Before **MLD** became popular, **compression pumping** was one of the main treatments for **lymphedema**. **Pumps** are available that can produce external pressures high enough to push fluid from a limb (arm or leg) and yet gentle enough not to cause harm to tissue. The use of the **compression pumps** is controversial by those who believe that **MLD** is the best way to gently encourage the fluid to move out the limb. There is concern that the **pump** pressures are too high and may be traumatic to the tissue. There are newer **pumps** that may actually mimic the motions of **MLD** and are programmed to do so. In addition, the ability to manipulate every phase of the **pumping** sessions by adjusting the pressures and timing sequences make **compression pumping** attractive to those with busy lifestyles. The **pumping** devices are compact enough to carry and use during travel allowing patients with **lymphedema** to have a lifestyle not dependent upon a therapist or family member to help handle their daily routine of **edema** reduction. Patient's experiencing this type of **pumping** session which lasts about 1 hour per limb report that the motion is quite tolerable and evidence has suggested that it is very effective. Your therapist will help you make decisions about which is the best approach for you.

The **maintenance phase** of therapy begins after the affected limb is reduced as much as can be expected (determined by daily measurements of the limb by the therapist). The goal of **maintenance therapy** is to prevent the limb from swelling again. **Compression** garments play a critical role in preventing swelling from happening again. The patient is given a prescription for a **compressions stocking**. It is important that the stocking fit the size of your limb with all the **edema** out since fluid will again appear if the stocking is too big. An experienced fitter should measure your limb. You should not stop wearing your **compression wrapping** until the stocking is available. The stocking should fit snug but should not be painful. You should put the stocking on as soon as you awake from sleep and preferably before you get out bed in the morning and should only take it off when you are ready to go to bed. **Compression stockings** can have pressures of 20-30 mmHg (for arm **lymphedema**) and 30-40 mmHg or even up to a custom made stocking providing 50 mmHG of pressure for lower extremity **lymphedema**. The pressure is

determined by the severity of the **lymphedema** and the previous success with lower pressure **compression stockings**. The stocking should be replaced every 4-6 months because they wear out. The patient should have 2 garments at the same time, so that one can be worn while the other is being washed. Always put the garment on using rubber or similar gloves to avoid putting a hole into the garment. The garment loses its **compression** once a hole gets in it. If 10-15 pounds or more are lost during the **maintenance phase**, a re-measured stocking is likely needed since the stocking will not being fitting well. If the affected limb begins to get larger despite your wearing the compression garment you should contact your therapist. Re-measurement is needed every year to make sure that the fit is still correct. Sometimes there may be additional thickened skin and fluid that could be reduced out of the limb in the year following a reduction phase. This may happen as the body remodels itself. There might be consideration given to another reduction phase of treatment if this does occur.

In addition to this standard program, you doctor may have you use **compression wrapping** at night to help reduce swelling even further. It is very important to continue exercising for best decrease in swelling and to improve the general quality of life. Walking, swimming, and bicycling are all good exercises to strengthen your muscles, help you to maintain a healthy weight and also help to keep **edema** from returning the affected limb. You should always wear your **compression** garment or wrap when you exercise. Elevation of the affected limb or body part allows gravity to help in draining the fluid especially at the end of the day. Raising the affected limb higher than the level of the heart is the recommended height. Some patients need **compression pumping** or **MLD** periodically or at night to keep the limb size down. Skin care is as important in this phase of treatment as during the reduction phase. Additional hints to improving the chances of having the best results are to avoid extreme temperatures such as getting into hot tubs or saunas. These can allow dilation of **blood vessels** and fluid to come back into the leg. Having an antibiotic available to be used at the first sign of infection may help to decrease recurrent problems with skin and fatty tissue infections. There are not proven effective drugs to treat lymphedema.

Surgical Treatment for Lymphedema

A variety of surgical techniques have been proposed to treat patients with **lymphedema** especially when it has become too difficult to manage by medical therapy alone. The type of **lymphedema** may determine if a surgical treatment is possible for your condition. **Excisional operations** remove (cut out) excess tissue to help decrease the size of the limb. The most extensive **excisional operation** is the **Charles's procedure** which removes all the skin and subcutaneous tissue from the knee down to the ankle. This procedure may be done when there have been multiple infections in the skin and fatty tissue under the skin, skin sloughing or the patient is having difficulty in performing normal daily functions. Reducing the bulk of skin is usually helpful but this invasive procedure can have complications such as poor wound healing, prolonged hospitalization, long scars, changes in feeling, and swelling in the foot and ankle. A less extensive form of surgery to reduce **lymphedema** tissue is **liposuction**. **Lymphedema** tissue in time turns into fat which can not be helped by **compression** so **liposuction** does this for the

body. In rare cases and when there are nicely open **lymph vessels** above and below an area of damage, **lymphatic reconstruction** or direct reconnections of the **lymph vessels** to each other or a vein is possible. These are called **lymphovenous anastomosis** or **lymphatic grafting** and use microsurgery. It is noted that this procedure is most useful when done early in the course of **lymphedema**, before subcutaneous fibrosis occurs and **lymphatic vessel** sclerosis or hardening begins and the **lymphatic vessels** still have the ability to contract. It has been done in patients with surgical damage to **lymph vessels** (such as after breast cancer surgery), **chylus reflux**, and in people affected with **filariasis** who often have very large **lymph vessels**. It is very delicate work working with very small vessels so it is done only in very select cases and the results are not well known.

Commonly asked questions

What is lymphedema?

Lymphedema is swelling in the leg, arm or other parts of the body due to the body's inability to effectively remove **lymph** fluid (which contains proteins, fatty acids, waste from cell feeding and bacteria that enter the body).

What can cause lymphedema?

Lymphedema can happen because the formation of the **lymph vessels** during maturation (as a preborn baby) was less than adequate (called **primary lymphedema**) or damage (injury) has occurred to the normally formed **lymph channels** and **lymph nodes** during surgery, from infection, from radiation treatment or other causes. It can also occur from too much **lymph** fluid being made such as can happen when **venous disease** is present. Because the **lymphedema** caused by damage or overproduction is the result of something other than a problem with how the **lymph vessels** matured it is called **secondary lymphedema**.

What can I do if I have lymphedema?

Your doctor can make sure that you actually have **lymphedema**. If you do and there is no infectious or other acute medical problem that must be dealt with, then external **compression** treatments are begun to decrease the swelling and to keep it down. This may involve external **massage**, **compression pumping**, **compression wraps** and eventually **compression stockings**. How well you do will be determined by how faithfully you use the treatments prescribed.

Is surgery for lymphedema a good option?

Surgery for **lymphedema** is really a last option for only a select group of patients. Medical treatment is the main stay of treatment and surgery is only considered if this fails. An in-depth discussion of this option with a vascular surgeon would be the best way to find out if you are a candidate for surgery.